# Neurosurgery Case Review: Questions and Answers

Second Edition

Remi Nader
Abdulrahman J. Sabbagh
Samer K. Elbabaa
Hosam Al-Jehani
Jaime Gasco
Cristian Gragnaniello

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# Neurosurgery Case Review: Questions and Answers

**2nd Edition**

**Remi Nader, MD, CM, FRCSC, FACS, FAANS**
Founder and President
Texas Center for Neurosciences and International Center for Neuroscience
Houston, Texas;
Adjunct Clinical Professor
Department of Neurosurgery
University of Texas Medical Branch
Galveston, Texas

**Abdulrahman J. Sabbagh, MBChB, FRCSC**
Assistant Professor and Consultant Neurosurgeon, Epilepsy Surgeon, and Pediatric Neurosurgeon
Neurosurgery Section;
Assistant Chairman for Research and Higher Education
Department of Surgery
College of Medicine;
Head of Research and Development
Clinical Skill and Simulation Center
King Abdulaziz University
Jeddah, Saudi Arabia

**Samer K. Elbabaa, MD, FAANS, FAAP, FACS**
Chief of Pediatric Neurosurgery and Director
Leon Pediatric Neuroscience Center of Excellence
Arnold Palmer Hospital for Children;
Professor of Neurosurgery
College of Medicine
University of Central Florida
Orlando, Florida

**Hosam Al-Jehani, MSc, FRCSC**
Assistant Professor and Consultant
Department of Neurosurgery, Interventional Neuroradiology, and Neurocritical Care
King Fahd Hospital of the University
Imam Abdulrahman Bin Faisal University
Alkhobar, Saudi Arabia;
Director
Neurosciences service line
EP-1 cluster
Dammam, Saudi Arabia

**Jaime Gasco, MD, FAANS, FEBNS**
Neurosurgeon
Department of Neurosurgery
University Medical Center of El Paso
El Paso, Texas

**Cristian Gragnaniello, MD, PhD, MSurg, MAdvSurg, FICS**
Assistant Professor of Neurological Surgery
Department of Neurological Surgery
University of Illinois at Chicago
Chicago, Illinois

**Section Editor (Intracranial Pathology: Tumors)**
**Hussam Abou-Al-Shaar, MD**
Neurosurgery Resident
Department of Neurological Surgery
University of Pittsburgh Medical Center
Pittsburgh, Pennsylvania

760 illustrations

Thieme
New York • Stuttgart • Delhi • Rio de Janeiro

**Library of Congress Cataloging-in-Publication Data** is available from the publisher

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Thieme Medical Publishers New York
333 Seventh Avenue
New York, New York 10001 USA
+1 800 782 3488
customerservice@thieme.com

Georg Thieme Verlag KG
Rüdigerstrasse 14, 70469 Stuttgart, Germany
+49 [0]711 8931 421, customerservice@thieme.de

Thieme Publishers Delhi
A-12, Second Floor, Sector-2, Noida-201301
Uttar Pradesh, India
+91 120 45 566 00, customerservice@thieme.in

Thieme Publishers Rio de Janeiro,
Thieme Publicações Ltda.
Edifício Rodolpho de Paoli, 25º andar
Av. Nilo Peçanha, 50 – Sala 2508,
Rio de Janeiro 20020-906 Brasil
+55 21 3172-2297

Cover design: Thieme Publishing Group
Typesetting by DiTech Process Solutions, India

Printed in USA by King Printing Company, Inc. 5 4 3 2 1

ISBN 978-1-62623-198-6

Also available as an e-book:
eISBN 978-1-62623-199-3

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This book is dedicated to...

All neurosurgery students and trainees around the world whom we ask that they do their very best to master neurosurgical knowledge and skills to help their patients.

-Editors

To my wife Irish and my parents Joseph and Eleonore.

-Remi Nader

Dedicated to my parents, my better half Alaa, our children, and my siblings

-Abdulrahman J. Sabbagh

To my wife Dina, for her everlasting patience and unwavering support, and to my children Kamal and Natalie, for touching my heart everyday and making it all worthwhile!
To all my pediatric neurosurgical patients I have had the honor to serve, for teaching me resilience and making the impossible possible everyday!

-Samer K. Elbabaa

To my wife Khuloud and my daughters Dana and Jumana for their unconditional support. To my parents Nahla and Maher for instilling in me the principle of endurance.
To my neurosurgical residents and fellows, for making every day a learning experience. And finally to my neurosurgical patients, from whom we garner our experience based on mutual trust and respect.

-Hosam Al-Jehani

To my wife Lina, brother Luis, and parents Vicente and Maria Teresa. To them I owe everything.

-Jaime Gasco

To my mother Anna and my wife Katie, the steady forces in my life that kept me going when I could have stopped.
To my Boss, Sergey, who taught me more than just technical wonders.

-Cristian Gragnaniello

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# Foreword

With today's rapid expansion of medical technology and clinical information, it is essential that we are on top of the fundamental knowledge in our field. To this end, Dr. Nader [and Associates] have compiled a remarkable collection of cases that highlight a variety of clinical entities and syndromes that help in this task.

The cases in this book cover the majority of topics of importance and interest to a junior neurosurgeon. As such, a careful review of all these cases would be particularly helpful in preparing for the in-training written board examination, as well as for the oral board examination. In fact, this comprehensive case book serves as a reminder of all the clinical and diagnostic entities that should be reviewed prior to taking these examinations. Very few publications exist that help accomplish this goal. This book therefore helps fill this void and provides a well thought-out outline of major neurosurgical topics and a high-quality collection of cases to review and study. The authors should be commended for their efforts.

*Raymond Sawaya, MD*
*Professor and Chairman*
*Department of Neurosurgery*
*The University of Texas M. D. Anderson Cancer Center*
*Houston, Texas*

# Preface

On being approached by the publisher, Thieme, to write a second edition of *Neurosurgery Case Review: Questions and Answers*, we were very surprised and excited. After being told that the first edition was one of their bestsellers among the neurosurgery textbooks, we feel greatly rewarded to have had such an impact on the contribution toward training of future neurosurgeons as well as recent graduates taking the board. Little did we know about the challenges that this project reserved for us as we took on the arduous task.

It has been a special challenge to coordinate the writings, updates, and revisions of the multiple chapters that make the text as well as the logistical organization of the multiple editors and section editors. Each one of us having a busy practice at sometimes opposite ends of the globe, we have more than once had to step back to the drawing board and try to reorganize the project before moving forward.

Although it may seem to the reader that a lot of material is repeated and therefore there is much less work in the creation of a second edition—this is not the case at all. We were surprised to find that we would have to review every single chapter and go through the laborious decision of resolving whether or not to keep or discard a chapter and also determine which topics need to be added. In order to do that, one has to have a thorough understanding of not only the depths, but the breadth of the field of neurosurgery. Currently, with specializations and sub-specializations in neurosurgery, we are more and more focused on a particular aspect of the field and we lose perception and vision about the global picture in neurosurgery. This is especially true for us, after having practiced neurosurgery for more than a decade now.

We have tried to capture the essence of the first edition; presenting a set of cases that present special challenges to the aspiring neurosurgeon and current practitioner. We would like to enrich their breadth of practice. To do so we have recruited more editors and authors to contribute new cases with more contemporary approaches to management. Conditions such as stroke, requiring endovascular treatment, are now required to be addressed by the neurosurgeons both at a didactic and a practical standpoint. Furthermore, there have been global advances in general neurosurgery, especially spine surgery, which we have also captured in this textbook. Neurosurgeons are now treating conditions such as scoliosis, sacroiliitis, and other ailments at the frontier of neurosurgery, as it overlaps with neurology, interventional radiology, orthopedic surgery, and pain management. We have covered aspects of the junction of these aforementioned disciplines via selected cases.

We have retained the spirit of the first edition, which includes focusing on the format of testing for the examinations of American Board Neurological Surgery and the Royal College of Physicians and Surgeons of Canada oral board examinations. In addition, we have also tried to increase our case numbers while eliminating superfluous cases. We have aimed to maintain the uniqueness of each case, showcasing distinct judgment. The art of practicing neurosurgery and individual authors' opinions are reflected through each particular case.

Now, with close to 100 contributors and authors from multiple countries and continents spanning the Americas, Europe, Africa, and Asia, we believe to have captured an excellent cross-section of neurosurgery as it is practiced today in the world, and more particularly with the North American standards.

*All chapters in this book are based on actual cases encountered by the primary or senior authors during the scope of their practices, and so are the complications and challenges discussed in each case. Any resemblance with cases or questions presented in formal examinations (such as national board examinations or licensing examinations) or with cases discussed in organized review courses, is purely coincidental.*

We hope you enjoy reading this text as much as we did preparing it.

*Remi Nader, MD, CM, FRCSC, FACS, FAANS*
*Cristian Gragnaniello, MD, PhD, MSurg, MAdvSurg, FICS*

# Contributors

**Ahmed T. Abdelmoity, MD**
Assistant Professor
Departments of Pediatrics and Neurology
Director of Department of Neurophysiology
Director of the Comprehensive Epilepsy Program
University of Missouri at Kansas City
University of Kansas
Children's Mercy Hospitals and Clinics
Kansas City, Missouri

**Hussam Abou-Al-Shaar, MD**
Neurosurgery Resident
Department of Neurological Surgery
University of Pittsburgh Medical Center
Pittsburgh, Pennsylvania

**Bassam M. J. Addas, MBChB, FRCSC**
Associate Professor
Division of Neurosurgery
Department of Surgery
King Abdulaziz University Hospital
Jeddah, Saudi Arabia

**Anup Aggarwal, MCh (Neurosurgery)**
Neurosurgeon
Texas Center for Neurosciences
Houston, Texas

**Charles B. Agbi, MD, FRCSC, FACS**
Associate Professor of Surgery and Neurosurgeon
Department of Surgery
The Ottawa Hospital
University of Ottawa
Ottawa, Canada

**Maqsood Ahmad, MMed (Neurosurgery)**
Senior Consultant Pediatric Neurosurgeon
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Abdulrazag Ajlan, MD**
Assistant Professor of Neurosurgery
Department of Surgery
College Of Medicine
King Saud Medical City
Riyadh City, Riyadh, Saudi Arabia

**Ahmed Alaqeel, MD**
Resident
Division of Neurosurgery
University of Calgary
Alberta, Canada

**Abdulmajeed Alahmari, MBBS**
Clinical Spinal fellow
Spinal Division
Department of Trauma and Orthopedics
University Hospitals of North Midlands
Staffordshire, United Kingdom

**Ayman Abdullah Albanyan, FRCS(C)**
Head
Pediatric Neurosurgery Section
Consultant Pediatric Neurosurgeon
Department of Neurosurgery
Neuroscience Center
King Fahd Medical City
Riyadh, Saudi Arabia

**Abdulrahman Albakr, MD**
Neurosurgery Resident
Department of Neurosurgery
University of Calgary
Alberta, Canada

**Homoud AlDahash, MBBS, MIM**
Program Director Neurosurgery
Residency Training program;
Consultant Neurosurgeon
King Faisal Specialist Hospital and Research Center
Riyadh, Saudi Arabia

**Rihaf Algain, MBBS**
Resident
Department of Internal Medicine
King Abdulaziz Medical City
National Guard Health Affairs
Riyadh, Saudi Arabia

**Almunder Algird, MD, FRCSC**
Assistant Professor
Department of Surgery
Division of Neurosurgery
McMaster University
Hamilton Health General Hospital
Ontario, Canada

**Mohammed Alghamdi, MD**
Neurosurgery Resident
Division of Neurosurgery
Department of Surgery
University of Ottawa
Ottawa, Canada

**Hosam Al-Jehani, MSc, FRCSC**
Assistant Professor and Consultant
Department of Neurosurgery, Interventional Neuroradiology, and Neurocritical Care
King Fahd Hospital of the university
Imam Abdulrahman Bin Faisal University
Alkhobar, Saudi Arabia;
Director
Neurosciences service line
EP-1 cluster
Dammam, Saudi Arabia

**Fahad AlKherayf, MD, MSc, CIP, FRCSC**
Assistant Professor
Division of Neurosurgery
Department of Surgery;
Assistant Professor
Department of Otolaryngology—Head and Neck Surgery;
Director of Minimally Invasive and Skull Base Surgery Fellowship
Director of Neurosurgery Residency Program
University of Ottawa
Ottawa, Canada

**Afnan Uthman Alkhotani, MD**
Neurosurgery Specialist
Department of Neuroscience
Section of Neurosurgery
King Faisal Specialist Hospital and Research Center
Jeddah, Saudi Arabia

**Ossama Al-Mefty, MD, FACS**
Director of Skull Base Surgery
Department of Neurosurgery
Brigham and Women's Hospital
Harvard Medical School
Boston, Massachusetts

**Mohammed Almekhlafi, MD, MSc, FRCPC**
Assisstant Professor
Department of Internal Medicine
King Abdulaziz University
Jeddah, Saudi Arabia

**Mohammad Almubaslat, MD, FAANS, FACS**
Neurological Surgery
Advanced Brain and Spine Institute
Mandeville, Louisiana

**Khalid Almusrea, MD, FRCS(C)**
Consultant
Department of Spine and Neurosurgery;
Director
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Soha Abdu M. Alomar, MD, MPH, FRCSC**
Assistant Professor and Consultant Neurosurgeon
Division of Neurosurgery
Department of Surgery
King Abdulaziz University
Faculty of Medicine
Jeddah, Saudi Arabia

**Naif M. Alotaibi, MD, MSc, FRCSC**
Clinical Fellow-Cerebrovascular Section
Department of Neurosurgery
Massachusetts General Hospital
Boston, Massachusetts

**Fahad Eid Alotaibi, MD, FRCSC**
Pediatric Neurosurgery
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Faisal Al-Otaibi, MD**
Associate Professor of Neurological Surgery, Epilepsy Surgery, Stereotactic and Functional Neurosurgery
Division of Neurological Surgery
Department of Neuroscience
King Faisal Specialist Hospital and Research Center
Alfaisal University
Riyadh, Saudi Arabia

**Ashwag Al-Qurashi, MCh**
Senior Registrar
Division of Neurosurgery
Department of Surgery
King Abdulaziz University Hospital
Jeddah, Saudi Arabia

**Mariam Alrashid, MD**
Resident
Department of Neurosurgery
McGill University
Montreal, Canada

**Nabeel S. Alshafai, MD, FRCSC, FEANS**
Assistant Professor of Neurosurgery and Spine
Department of Neurosurgery
University of Antwerp
Antwerp, Belgium

**Mohammad Misfer Alshardan, MD**
Neurosurgery Resident
Division of Neurosurgery
University of Ottawa
Ottawa, Canada

**Ibrahim Althubaiti, MD**
Division of Neurosurgery
Department of Neurosciences
King Faisal Specialist Hospital and Research Center
Jeddah, Saudi Arabia

**Abdulrahman Y. Alturki, MSc, FRCSC**
Subspecialty Consultant
Vascular Neurosurgery, Endovascular Neurosurgery and Neurocritical Care
Department of Adult Neurosurgery;
Section head
Neurocritical Care Unit
National Neurosciences Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Shaymaa Al-Umran, MD**
Department Of Neurosurgery
Dammam Medical Complex
Dammam, Saudi Arabia

**Jorge E. Alvernia, MD, FAANS**
Associated Faculty
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**Ali Alwadei, MD**
Demonstrator (Resident)
Department of Neurosurgery
Imam Abdulrahman Bin Faisal University
Dammam, Eastern Province, Saudi Arabia

**Mahmoud AlYamany, MD, MHA, FRCSC, FAANS**
Neurovascular and Skullbase Neurosurgeon
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Ahmed Alzahrani, MD**
Consultant
Department of Spine and Neurosurgery
Security Forces Hospital
Riyadh, Saudi Arabia

**Jeffery Atkinson, MD, RCS(C)**
Assistant Professor
Department of Neurosurgery
McGill University
Montreal Children's Hospital
Quebec, Canada

**Mohammad A. Aziz-Sultan, MD**
Associate Professor of Neurosurgery
Department of Neurosurgery
Brigham and Women's Hospital
Boston, Massachusetts

**Saleh S. Baeesa, MD, FRCSC**
Head
Division of Neurosurgery
Department of Surgery
Faculty of Medicine
King Abdulaziz University
Jeddah, Saudi Arabia

**Mohammed Saeed Bafaqeeh, MD**
Assistant Professor of Neurosurgery
Department of Neurosurgery
National Neuroscience Institute
Kind Fahad Medical City
Riyadh, Saudi Arabia

**Christopher D. Baggott, MD**
Neurosurgeon
SSM Health St. Mary's Hospital
Madison, Wisconsin

**Michele Bailo, MD**
Neurosurgeon
Department of Neurosurgery and Gamma Knife Radiosurgery
San Raffaele Scientific Institute
Vita-Salute University
Milan, Italy

**Khalid Bajunaid, MSc, MMgmt, FRCSC**
Assistant Professor of Neurosurgery, Neurointensivist, Neurovascular Neurosurgeon, and Neuro-Interventionist
Department of Surgery
Faculty of Medicine
University of Jeddah
Jeddah, Saudi Arabia

**Layla Batarfi, MD**
Department of Neurosurgery
King Fahd University Hospital
Al Khobar, Saudi Arabia

**Da'Marcus Baymon, MD**
Emergency Medicine Resident
Boston Children's Hospital
Boston, Massachusetts

**Salah Baz, MD**
Consultant Neurologist
Department of Neurosciences
Section of Adult Neurology
King Faisal Specialist Hospital and Research Center
Jeddah, Saudi Arabia

**Han Zhuang Beh, MD**
Resident, PGY-6 (Integrated)
University of Texas Medical Branch
Galveston, Texas

**André Beer-Furlan, MD, PhD**
Resident Physician
Department of Neurological Surgery
Rush University
Chicago, Illinois

**Edward C. Benzel, MD**
Emeritus Chair
Department of Neurosurgery
Cleveland Clinic
Cleveland, Ohio

**Michel W. Bojanowski, MD, FRCSC**
Professor of Surgery
Division of Neurosurgery
University of Montreal;
Neurosurgeon
University of Montreal Medical Center
Quebec, Canada

**Perry S. Bradford, MD**
Plastic Surgery Resident
Department of Plastic Surgery
University of Virginia Health System
Charlottesville, Virginia

**Daniel Branch, MD**
Resident, PGY-6
Department of Neurosurgery
University of Texas Medical Branch School of Medicine
Galveston, Texas

**Frederick Boop, MD**
Professor
Department of Surgery
Division of Neurosurgery
University of Tennessee
Memphis, Tennessee

**Christian A. Bowers, MD**
Assistant Professor of Neurosurgery
Department of Neurosurgery
New York Medical College
New York, New York

**Richard Bucholz, MD, FACS**
Professor
Division of Neurological Surgery
Saint Louis University
St. Louis, Missouri

**Reem Bunyan, MD**
Consultant Neurologist and Neuroimmunologist
Neuroimmunology Program
Departments of Neurology and Neurophysiology
Chair of Neurophysiology
Neurosciences Center
Kind Fahd Medical City
Riyadh, Saudi Arabia

**Alwin Camacho, MD**
Assistant Clinical Professor
Department of Radiology
The University of Texas Medical Branch
Galveston, Texas

**Anthony J. Caputy, MD, FACS**
Chairman
Department of Neurological Surgery
The George Washington University
Washington, District of Columbia

**Ricardo L. Carrau, MD, MBA**
Professor and Lynne Shepard Jones Chair in Head and Neck Oncology
Department of Otolaryngology—Head and Neck Surgery and Department of Neurological Surgery;
Director of the Comprehensive Skull Base Surgery Program
The Ohio State University
Wexner Medical Center
Columbus, Ohio

**Pratap Chand, MD**
Professor of Neurology and Psychiatry
Division of Neurological Surgery
Saint Louis University
St. Louis, Missouri

**Stephano Chang, BSc. MD (PGY5)**
Resident
Division of Neurosurgery
Department of Surgery
University of Toronto
Toronto, Canada

**Alvin Chan, MD**
Resident Physician
Department of Pediatrics
University of California
Irvine, California

**Anthony M. T. Chau, MAppAnatDiss, MSurg**
Neurosurgery Registrar
Department of Neurosurgery
Sir Charles Gairdner Hospital
Perth, Australia

**Claude-Edouard Châtillon, MSc, MD, FRCSC**
Assistant professor
Department of Neurosurgery
University of Montreal
Quebec, Canada

**Ennio Antonio Chiocca, MD, PhD, FAANS**
Harvey W. Cushing Professor of Neurosurgery
Harvard Medical School;
Neurosurgeon-in-Chief and Chairman
Department of Neurosurgery
Brigham and Women's Hospital
Boston, Massachusetts

**William T. Couldwell, MD, PhD**
Professor and Chair
Department of Neurosurgery
University of Utah
Salt Lake City, Utah

**Franco DeMonte, MD, FRCSC, FACS**
Professor
Departments of Neurosurgery and Head and Neck Surgery;
Mary Beth Pawelek Chair in Neurosurgery
The University of Texas MD Anderson Cancer Center
Houston, Texas

**Badih Daou, MD**
Neurosurgery Resident
Department of Neurosurgery
University of Michigan
Ann Arbor, Michigan

**Sohum Desai, MD**
Neurosurgeon
Doctors Hospital at Renaissance
Harlingen, Texas

**Randall C. Edgell, MD, FSVIN**
Professor
Department of Neurology;
Director, Souers Stroke Institute;
Director, Neurointerventional Services;
Director, Vascular and Neurointerventional Fellowships;
Saint Louis University
St. Louis, Missouri

**Turki Elarjani, MD**
Neurosurgery Resident
Department of Neurosciences
Section of Neurosurgery
King Faisal Specialist Hospital and Research Center
Riyadh, Saudi Arabia

**Samer K. Elbabaa, MD, FAANS, FAAP, FACS**
Chief of Pediatric Neurosurgery and Director
Leon Pediatric Neuroscience Center of Excellence
Arnold Palmer Hospital for Children;
Professor of Neurosurgery
College of Medicine
University of Central Florida
Orlando, Florida

**Nicholas J. Erickson, MD**
Resident, PGY-3
Department of Neurological Surgery
University of Alabama at Birmingham
Birmingham, Alabama

**Isabella Esposito, MD**
Neurosurgeon
Department of Neurosurgery
Colli Hospital—Vincenzo Monaldi Hospital
Naples, Italy

**Domenic P. Esposito, MD, FACS**
Retired Associate Professor
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**H. Francis Farhadi, MD, PhD, FRCS(C), FAANS**
Chief,
Division of Degenerative and Deformity Spinal Surgery;
Director, Spinal Surgery Fellowship Program
Department of Neurological Surgery
The Ohio State University Wexner Medical Center
Columbus, Ohio

**Jean-Pierre Farmer, MD, CM, FRCS(C)**
Professor
Department of Neurosurgery
McGill University
Montreal Children's Hospital
Quebec, Canada

**Faisal Abdulhamid Farrash, SABNS, MIM**
Consultant
Department of Neurosurgery;
Assistant Professor
Department of Neuroscience
King Faisal Specialist Hospital and Research Center
Al Faisal University
Riyadh, Saudi Arabia

**Jared Fridley, MD**
Director
Spinal Surgical Outcomes Laboratory
Neurosurgery Foundation—Lifespan Physician Group
Providence, Rhode Island

**Aaron S. Gaekwad, BMBS, B (Pharma)**
Senior Medical Registrar and Adult Medicine Physician Trainee
Division of Medicine and Sub-specialty
Lyell McEwin Hospital
Northern Adelaide Local Health Network
Elizabeth Vale, Australia

**Filippo Gagliardi, MD, PhD**
Neurosurgeon
Department of Neurosurgery and Gamma Knife Radiosurgery
San Raffaele Scientific Institute
Vita-Salute University
Milan, Italy

**Gary L. Gallia, MD, PhD**
Surgical Director
Department of Neurosurgery
Pituitary Center
Johns Hopkins University School of Medicine
Baltimore, Maryland

**Alberto L. Gallotti, MD**
Resident
Department of Neurosurgery and Gamma Knife Radiosurgery
San Raffaele Scientific Institute
Vita-Salute University
Milan, Italy

**Jaime Gasco, MD, FAANS, FEBNS**
Neurosurgeon
Department of Neurosurgery
University Medical Center of El Paso
El Paso, Texas

**Lorenzo Genitori, MD**
Neurosurgeon
Department of Neurosurgery
Anna Meyer Pediatric Hospital
Florence, Italy

**Fred Gentili, MD, MSc, FRCSC**
Professor of Surgery and Otolaryngology
Faculty of Medicine
University of Toronto;
Director of Surgical Education
University Health Network;
Associate Staff
Mount Sinai Hospital
Toronto, Ontario

**Frank Gerold, MD**
Assistant Professor
Department of Orthopedic Surgery
University of Texas Rio Grande Valley
Edinburg, Texas

**Ehtesham Ghani, FCPS (Neurosurgery)**
Assistant Consultant
Department of Neurosurgery
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Diana Ghinda, MD**
Neurosurgery Resident
Division of Neurosurgery
Department of Surgery
University of Ottawa
Ottawa, Canada

**Paul W. Gidley, MD, FACS**
Professor of Otology, Neurotology, and Skull Base Surgery
Department of Head and Neck Surgery
The University of Texas MD Anderson Cancer Center
Houston, Texas

**Brian Gill, MD**
Resident Physician
Department of Neurological Surgery
Columbia University
New York, New York

**Flavio Giordano, MD**
Consultant Neurosurgeon
Department of Neurosurgery
Anna Meyer Pediatric Hospital
Florence, Italy

**Jason S. Goldberg, MD**
Ophthalmology Resident, PGY-4
Department of Ophthalmology
Georgetown University Hospital
Washington Hospital Center
Washington, District Of Columbia

**Pablo González-López, MD, PhD**
Associate Professor
Department of Neurosurgery
General University Hospital of Alicante
Alicante, Spain

**Cristian Gragnaniello, MD, PhD, MSurg, MAdvSurg, FICS**
Assistant Professor of Neurological Surgery
Department of Neurological Surgery
University of Illinois at Chicago
Chicago, Illino

**Eldad J. Hadar, MD**
Neurosurgeon
Department of Neurosurgery
University of North Carolina
Chapel Hill, North Carolina

**Mark G. Hamilton, MDCM, FRCSC, FAANS**
Professor of Neurosurgery
Department of Clinical Neurosciences
Division of Neurosurgery
University of Calgary
Alberta, Canada

**Kimberly Hamilton, MD**
Rheumatology specialist
NCH Healthcare Systems
Naples, Florida

**Amgad S. Hanna, MD**
Associate Professor
Department of Neurological Surgery
University of Wisconsin
Madison, Wisconsin

**Marguerite Harding, MBChB, MMed, FRACS and Masters of Surgical Education**
Neurosurgeon
Department of Neurosurgery
Flinders Medical Center
Bedford Park, Australia

**Hashem Al Hashemi, MD, FRCPC**
Physical Medicine and Rehabilitation Consultant
Division of Physical Medicine and Geriatric
Department of Medicine
King Abdulaziz Medical City
Riyadh, Saudi Arabia

**Roberto Rafael Herrera, MD**
Head of Neurosurgery
Department of Neurosurgery
Belgrano Adventist Clinic;
Board of Director
Walter Dandy Neurosurgical Society
Buenos Aires, Argentina

**Stephen J. Hentschel, MD**
Clinical Assistant Professor
Department of Surgery
University of British Columbia
Victorial General Hospital
Vancouver, Canada

**Joaquin Hidalgo, MD**
Neurosurgery Staff
Department of Neurosurgery
North Mississippi Health System
Tupelo, Mississippi

**Qasim Al Hinai, BSc, MD, MRCSI, FRCSC (Neurosurgery)**
Senior Consultant Neurosurgeon
Department of Neurosurgery
Khoula Hospital, Ministry of Health
Muscat, Oman

**Glenn C. Hunter, MD, FRCSC**
Professor of Surgery Emeritus
Department of Surgery
University of Arizona
Tucson, Arizona

**Daniel S. Ikeda, MD, FAANS**
Staff Neurosurgeon;
Chair of Surgery;
Assistant Professor of Surgery
Uniformed Services University of the Health Sciences
US Naval Hospital Okinawa
Ginowan, Japan

**Albert M. Isaacs, MD**
Resident
Division of Neurosurgery
University of Calgary
Alberta, Canada

**Jorge E. Isaza, MD**
Orthopedic Surgeon
Department of Neurosurgery
Baton Rouge General Medical Center
Baton Rouge, Louisiana

**Pascal M. Jabbour, MD**
Professor of Neurological Surgery
Chief Division of Neurovascular Surgery and Endovascular Neurosurgery
Thomas Jefferson University Hospital
Philadelphia, Pennsylvania

**Randy L. Jensen, MD, PhD**
Professor and Residency Program Director
Department of Neurosurgery
Clinical Neurosciences Center
University of Utah
Salt Lake City, Utah

**Andrew Jea, MD**
Pediatric Spine Surgeon and Neurosurgeon;
Director of the NeuroSpine program
Texas Children's Hospital
Houston, Texas

**Julius July, MD, PhD, IFAANS**
Associate Professor and Associate Dean for Clinical Education
Department of Neurosurgery
Faculty of medicine
Universitas Pelita Harapan
Neuroscience Center Siloam Hospital Lippo Village
Banten, Indonesia

**Imad N. Kanaan, MD, FACS, FRCS (Edinburgh)**
Professor and Chairman
Department of Neurosciences
King Faisal Specialist Hospital and Research Center
Alfaisal University College of Medicine
Riyadh, Saudi Arabia

**Amin B. Kassam, MD**
Chief Scientific Strategist
Advocate Aurora Health Care;
Vice-President
Aurora Neuroscience Innovation Institute;
Chairman
Department of Neurosurgery
Aurora St. Luke's Medical Center
Milwaukee, Wisconsin

**Robert F. Keating, MD**
Professor and Chair
Department of Neurosurgery
Children's National Medical Center
George Washington University School of Medicine
Washington, District of Columbia

**Patrick Kim, AB, MD**
Neurosurgery Resident
Department of Neurosurgery
University of Mississippi
Jackson, Mississippi

**Denis Klironomos, MD, FRCS(S)**
St. Luke's Neurology Associates
Bethlehem, Pennsylvania

**Kathleen E. Knudson, MD**
Pediatric Neurosurgery Fellow
Department of Neurological Surgery
Cincinnati Children's Hospital Medical Center
Cincinnati, Ohio

**Sunil Kukreja, MBBS**
Orthopedic Spine Surgeon
Datta Endoscopic Back Surgery and Pain Center
Saddle Brook, New Jersey

**Mohamed A. Labib, MDCM, FRCSC**
Neurosurgery Resident
Department of Neurosurgery
Barrow Neurological Institute
Phoenix Arizona

**Michel Lacroix, PhD, FRCS(C), FACS**
Director of Neurosurgery and Director of Brain and Spine Tumor Institute
Department of Neurosurgery
Geisinger Wyoming Valley Medical Center
Wilkes-Barre, Pennsylvania

**Ahmad I. Lary, MD, FRCS(C)**
Director of Neuro-oncology Program
Department of Neurosurgery
Neuroscience Center
King Fahd Medical City
Riyadh, Saudi Arabia

**Jonathon Lebovitz, MD**
Neurosurgeon
Neurosurgery Associates of Southwestern Connecticut
Philadelphia, Pennsylvania

**José Luis Ledesma, MD**
Neurosurgeon
Department of Neurosurgery
Belgrano Adventist Clinic
Buenos Aires, Argentina

**Joung H. Lee, MD**
Professor
Head of Skull Base Surgery
Brain Tumor and Neuro-oncology Center
Neurological Institute
Cleveland Clinic
Cleveland, Ohio

**Hubert Lee, MD, MSc**
Neurosurgery Resident
Division of Neurosurgery
Department of Surgery
University of Ottawa
Ottawa, Canada

**Alexander Y. Lin, MD, FACS**
Professor and Chief of Pediatric Plastic Surgery;
Director
St. Louis Cleft-Craniofacial Center
SSM Health Cardinal Glennon Children's Hospital
Saint Louis University;
Wolff Endowed Chair in Craniofacial, Maxillofacial, and Pediatric Plastic Surgery
Division of Plastic Surgery
Saint Louis University School of Medicine
Saint Louis, Missouri

**Zachary N. Litvack, MD**
Director, Skull Base and Minimally Invasive Neurosurgery
Department of Neurosurgery
Swedish Neuroscience Institute
Seattle, Washington

**Joshua Loewenstein, MD**
Resident Physician
Department of Neurosurgery
University of North Carolina
Chapel Hill, North Carolina

**Ali Luqman, MD**
Neurosurgery Resident
Department of Neurosurgery
University of Mississippi
Jackson, Mississippi

**Gustavo D. Luzardo, MD**
Associate Professor of Neurosurgery
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**Rolando Del Maestro, MD, PhD, FRCS(C), FACS**
Professor
Department of Neurology and Neurosurgery
McGill University
Montreal Neurological Institute
Quebec, Canada

**Mark A. Mahan, MD**
Assistant Professor
Department of Neurosurgery
University of Utah
Salt Lake City, Utah
**Monish Maharaj, MD**
Research Coordinator
Department of Neurosurgery
NeuroSpine Surgery Research Group
Sydney, Australia

**Ramez Malak, MD, MSc, FRCSC, FAANS**
Head
Department of Neurosurgery
Charles LeMoyne Hospital Greenfield Park
Quebec, Canada;
Professor
Department of Neurosurgery
University of Sherbrooke
Quebec, Canada

**Judith Marcoux, MD, MSc, FRCSC**
Neurosurgeon
Department of Neurosurgery
McGill University Health Center
Quebec, Canada

**Jared J. Marks, MD**
Instructor of Neurosurgery
Department of Neurosurgery
Johns Hopkins Medicine
Bethesda, Maryland

**Evan S. Marlin, MD**
Director of Endovascular Neurosurgery
St. Luke's University Health Network
St. Luke's Neurosurgical Associates
Bethlehem, Pennsylvania

**Rory Mayer, MD**
Fellow, Neurological Surgery
UCSF Weill Institute for Neurosciences
School of Medicine
Francisco, California

**Nancy McLaughlin, MD, PhD**
Department Family and Emergency Medicine
University of Montreal
Montreal, Quebec, Canada

**Peter J. Mews, MMedSc, FRACS**
Consultant Neurosurgeon and Endovascular Neurosurgeon
Department of Neurosurgery
The Canberra Hospital
Australian National University
Canberra, Australia

**Pietro Mortini, MD**
Chairman
Department of Neurosurgery and Gamma Knife Radiosurgery
San Raffaele Scientific Institute
Vita-Salute University
Milan, Italy

**Philippe Mercier, MD, PhD**
Assistant Professor
Division of Neurosurgery
Saint Louis University
St. Louis, Missouri

**Sandeep Mittal, MD, FRCSC, FACS**
Professor
Department of Neurosurgery
Virginia Tech Carilion School of Medicine;
Fralin Biomedical Research Institute;
Neurosurgeon
Carilion Clinic
Roanoke, Virginia

**Ralph J. Mobbs, BSc, MB, BS, MS, FRACS**
Chair
NeuroSpine Surgery Research Group;
Head
Department of Neurosurgery
Prince of Wales Private Hospital
Sydney, Australia

**José Luis Montes, FACS**
Former Director of Neurosurgery
Department of Neurosurgery, Neurology, and Oncology
Montreal Children Hospital
McGill University
Quebec, Canada;
Consultant Neurosurgeon
Medical School Autonomous University
San Luis Potosí, Mexico

**Ahmed Mohyeldin, MD**
Clinical Instructor
Department of Neurosurgery
Stanford University
Stanford, California

**Amal Mokeem, MD**
Consultant Clinical Neurophysiologist
Division of Clinical Neurophysiology
Department of Neuroscience
King Faisal Specialist Hospital and Research Center
Riyadh, Saudi Arabia

**Robert A. Moumdjian, MD**
Professor
Department of Neurosurgery
University of Montreal
Quebec, Canada

**Jeffrey P. Mullin, MD, MBA**
Assistant Professor
Department of Neurosurgery
University at Buffalo Neurosurgery
Buffalo, New York

**John S. Myseros, MD**
Professor and Vice Chief
Department of Neurosurgery
Children's National Hospital
Washington, District of Columbia

**Remi Nader, MD, CM, FRCSC, FACS, FAANS**
Founder and President
Texas Center for Neurosciences
International Center for Neuroscience
Houston, Texas;
Chairman
Department of Neurosurgery
United General Hospital
Houston, Texas;
Adjunct Clinical Professor
Department of Neurosurgery
University of Texas Medical Branch
Galveston, Texas

**Marc-Elie Nader, MD, CM, MSc, FRCSC**
Assistant Professor of Otology, Neurotology, and Skull Base Surgery
Department of Head and Neck Surgery
The University of Texas MD Anderson Cancer Center
Houston, Texas

**Abdul Haseeb Naeem, MD**
Chief Resident, PGY 6
Department of Clinical Neurological Sciences
London Health Sciences Center
Ontario, Canada

**Peter Nakaji, MD**
Professor and Chair
Department of Neurosurgery
University of Arizona College of Medicine;
Executive Director
Neurosciences Institute at Banner University Medical Center
Phoenix, Arizona

**Ivona Nemeiko, MD**
Neurosurgical Staff Specialist
Department of Neurosurgery
Aarhus University Hospital
Aarhus, Denmark

**Weston T. Northam, MD**
Neurosurgeon
Department of Neurosurgery
University of North Carolina
Chapel Hill, North Carolina

**Sami Obaid, MD, CM, FRCSC**
Neurosurgeon
Department of Neurosurgery
University of Montreal
Quebec, Canada

**Ibrahim Omeis, MD**
Neurosurgeon
Department of Neurosurgery
Baylor College of Medicine
Houston, Texas

**Edgar Gerardo Ordóñez-Rubiano, MD**
Neurosurgeon and Assistant Professor
Department of Neurological Surgery
Foundation University of Health Sciences
Bogotá, Colombia

**Juan Ortega-Barnett, MD**
Assistant Professor
Department of Neurosurgery
Dell Seton Medical Center
The University of Texas
University of Texas Medical Branch
Shenandoah, Texas

**Achal Patel, MD**
Neurosurgeon
Doctors Hospital at Renaissance
Edinburg, Texas

**Vishal Patel, MD**
Assistant Professor of Neurology and Neurosurgery
Department of Neurosurgery
Emory University School of Medicine
Atlanta, Georgia

**Kamlesh B. Patel, MD, MSc**
Associate Professor of Surgery
Division of Plastic and Reconstructive Surgery
Washington University School of Medicine
St. Louis Children's Hospital
St. Louis, Missouri

**Justine Pearl, MD**
Practicing Neurosurgeon
Department of Neurosurgery
McLaren Regional Medical Center
Flint, Michigan

**Lissa Marie Peeling, BSc, MD, FRCSC**
Associate Professor of Surgery
Division of Neurosurgery
Department of Surgery
College of Medicine
University of Saskatchewan
Saskatoon, Canada

**Eddie Perkins, PhD**
Associate Professor
Department of Neurosurgery
University of Mississippi
Jackson, Mississippi

**Kevin Petrecca, MD, PhD, FRCS(C)**
Neurosurgeon, Associate Professor, and Chief
Department of Neurosurgery
McGill University
Montreal Neurological Institute
Montreal, Canada

**Kevin Phan, MD, MSc, MPhil**
Resident Medical Officer
Department of Neurosurgery
Prince of Wales Clinical School
NeuroSpine Surgery Research Group
Sydney, Australia

**Matthew Pierson, MD**
Cerebrovascular/Skull Base Fellow
Department of Neurosurgery
Saint Louis University
St. Louis, Missouri

**Daniel M. Prevedello, MD**
Professor
Department of Neurological Surgery
The Ohio State University
Columbus, Ohio

**Nazer Qureshi, MD, D. Stat, MSc, FAANS, FICS, FACS**
Chief of Neurosurgery
Robert Wood Johnson/Barnabas Health
Hamilton, New Jersey;
Neurosurgeon
Department of Neuro-oncology
Princeton Brain and Spine Care
Princeton, New Jersey

**Shaan M. Raza, MD, FAANS**
Associate Professor of Neurosurgery and Head and Neck Surgery
Department of Neurosurgery;
Vice Chairman—Education
Skull Base Program
The University of Texas MD Anderson Cancer Center
Houston, Texas

**Justin Reagan, MD**
Physician
Department of Neurosurgery
Medical University of South Carolina
Charleston, South Carolinas

**Franz L. Ricklefs, MD**
Research Fellow
Department of Neurosurgery
University of Hamburg
Hamburg, Germany

**Rudiger Von Ritschl, MD**
Assistant Professor
Department of Radiology
University of Texas Medical Branch at Galveston
Galveston, Texas

**Giuliana Rizzo, MD**
Anaesthesiologist
Department of Neuroanesthesia and Neurorianimation
Anna Meyer Pediatric Hospital
Florence, Italy

**Eric P. Roger, MD, FRSC**
Neurosurgeon
Department of Neurosurgery
Buffalo General Hospital
Buffalo, New York

**Hector P. Rojas, MD**
Neurosurgeon
Department of Neurosurgery
Belgrano Adventist Clinic
Buenos Aires, Argentina

**Stephen M. Russell, MD**
Assistant Professor
Department of Neurosurgery
NYU school of Medicine
New York, New York

**Gareth Rutter, MBBS**
Registrar
Department of Neurosurgery
Royal Adelaide Hospital
Adelaide City Center, Australia

**Abdulrahman J. Sabbagh, MBChB, FRCSC**
Assistant Professor and Consultant Neurosurgeon,
Epilepsy Surgeon, and Pediatric Neurosurgeon
Neurosurgery Section;
Assistant Chairman for Research and Higher Education
Department of Surgery
College of Medicine;
Head of Research and Development
Clinical Skill and Simulation Center
King Abdulaziz University
Jeddah, Saudi Arabia

**Burak Sade, MD**
Clinical Associate
Section of Skull Base Surgery
Brain Tumor and Neuro-oncology Center
Neurological Institute
Cleveland Clinic
Cleveland, Ohio

**Asem Salma, MD**
Attending (Consultant) and Neurosurgeon
Department of Neurosurgery
Mercy Health—St. Rita's Medical Center
Lima, Ohio

**Francisco Sanz, MD, PhD**
Neurosurgeon
Department of Neurosurgery
Belgrano Adventist Clinic
Buenos Aires, Argentina

**Adam Sauh Gee Wu, MD**
Neurosurgery Resident
Division of Neurosurgery
University of Saskatchewan
Royal University Hospital
Saskatchewan, Canada

**Brian Seaman, DO, FACOS**
Neurosurgeon
Riverside Methodist Hospital;
Clinical Assistant Professor of Neurosurgery
Department of Neurosurgery
Ohio University Heritage College of Osteopathic Medicine
Columbus, Ohio

**Anish Sen, MD**
Neurosurgeon
Department of Neurosurgery and Trauma
Loma Linda University Medical Center
Loma Linda, California

**Essam A. Al Shail, MD, IFAANS**
President
Arab Pediatric Neurosurgical Society;
Professor and Director of Pediatric Neurosurgery
Department of Neurosciences
King Faisal Specialist Hospital and Research Center
Al Faisal University;
Riyadh, Saudi Arabia

**Rahul Shah, MD**
Orthopedic surgeon
Inspira Medical Center-Elmer
Vineland, New Jersey

**Andrew Shaw, MD, FAANS**
Staff Neurosurgeon
Baptist Medical Center
Jacksonville, Florida

**Joseph A. Shehadi, MD, FRCSC, FAANS**
Clinical Professor
Department of Neurosurgery
Ohio University Heritage College of Osteopathic Medicine
Columbus, Ohio

**Bassem Yousef Sheikh, MD**
Vice Dean for Clinical Affairs
Faculty of Medicine
Taibah University
Al-Madinah, Saudi Arabia

**Donald C. Shields, MD, PhD, MBA**
Associate Professor of Neurosurgery
Department of Neurosurgery
George Washington University School of Medicine and Health Sciences
Washington, District of Columbia

**Dimitri Sigounas, MD**
Assistant Professor of Neurosurgery
Department of Neurosurgery
The George Washington University
Washington, District of Columbia

**David Sinclair, MD, FRCSC (Neurosurgery), CSPQ (Neurochirurgie)**
Clinical Professor in Neurological Surgery
Department of Neurology and Neurosurgery
The Montreal Neurological Institute
McGill University
Quebec, Canada

**Denis Sirhan, MD, FRCSC**
Neurosurgeon;
Director of Cerebrovascular Surgery;
Director of Skull Base Surgery;
Department of Neurology and Neurosurgery
The Montreal Neurological Institute (MNI) of McGill University
Quebec, Canada

**Alan Siu, MD**
Neurosurgeon
Department of Neurosurgery
Ventura County Medical Center
Ventura, California

**Hasel W. Slone, MD**
Radiologist
Department of Radiology
The Ohio State University Wexner Medical Center
Columbus, Ohio

**Andrew Smith, MD, PhD**
Assistant Professor and the Director of Research
Department of Radiology
University of Mississippi Medical Center
Jackson, Mississippi

**Luke G. F. Smith, MD**
Resident
Department of Neurological Surgery
The Ohio State University
Wexner Medical Center
Columbus, Ohio

**Matthew D. Smyth, MD**
Appoline Blair Professor of Neurological Surgery and Pediatrics
Department of Neurosurgery
Washington University
St. Louis Children’s Hospital
St. Louis, Missouri

**Lahbib A. Soualmi, PhD**
Consultant, Image Guided Neurosurgical Navigation;
Head, Neuronavigation and Intraoperative Surgical Imaging
National Neuroscience Institute
King Fahad Medical City
Riyadh, Saudi Arabia

**Christopher Evan Stewart, DO**
Neurology Specialist
Department of Neurosurgery
Billings Clinic
Billings, Montana

**Paul Steinbok, BSc, FRCSC**
Professor
Division of Neurosurgery
BC Children’s Hospital
Vancouver, Canada

**Alfio Spina, MD**
Neurosurgeon
Department of Neurosurgery and Gamma Knife Radiosurgery
San Raffaele Scientific Institute
Vita-Salute University
Milan, Italy

**Michael S. Taccone, MD**
Neurosurgery Resident
Division of Neurosurgery
Department of Surgery
University of Ottawa
Ottawa, Canada

**Robert L. Tiel, MD†**
Professor
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**Jason Tullis, MD**
Associate Professor of Neurosurgery
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**Mazda K. Turel, MCh (Neurosurgery)**
Consultant Neurosurgeon
Wockhardt Hospital;
Honorary Assistant Professor
Department of Neurosurgery
Grant Medical College and Sir JJ Groups of Hospitals
Mumbai, Maharashtra, India

**Unwar Ul-Haq, FCPS**
Assistant Consultant Neurosurgeon
Department Of Neurosciences
King Faisal Specialist Hospital and Research Center
Al Faisal University
Riyadh, Saudi Arabia

**Exequiel P. Verdier, MD**
Attending Staff Neurosurgeon
Department of Pediatric Neurosurgery
Children's Hospital of San Justo
Buenos Aires, Argentina

**Iván Verdú-Martínez, MD**
Neurosurgeon
Department of Neurosurgery
General University Hospital of Alicante
Alicante, Spain

**Terence Verla, MD**
Medical Resident
Department of Neurosurgery
Baylor College of Medicine
Houston, Texas

**Ananth K. Vellimana, MD**
Resident
Department of Neurological Surgery
Washington University School of Medicine
St. Louis, Missouri

**Erol Veznedaroglu, MD, FACS, FAANS, FAHA**
Director
Department of Neurosurgery
Drexel University College of Medicine
Philadelphia, Pennsylvania

**Eka Julianta Wahjoepramono, MD, PhD, IFAANS**
Professor, Chairman, and Dean
Department of Neurosurgery
Faculty of Medicine
Universitas Pelita Harapan
Neuroscience Center Siloam Hospital Lippo Village
Banten, Indonesia

**Kelsey A. Walsh, MD**
Resident, PGY-VII
Department of Neurosurgery
University of Mississippi Medical Center
Jackson, Mississippi

**Craig C. Weinkauf, MD, PhD**
Assistant Professor
Department of Surgery
University of Arizona
Tuscon, Arizona

**John Woulfe, MD, PhD, FRCPC**
Assistant Professor
Department of Pathology;
Associate Scientist
Neuroscience Program
University of Ottawa
Ottawa, Canada

**Christopher J. Winfree, MD, FAANS**
Assistant Professor of Neurological Surgery
Department of Neurological Surgery
Columbia University
New York, New York

**Jonathan Yun, MD**
Attending in Neurological Surgery
Department of Neurological Surgery
Valley Hospital
Ridgewood, New Jersey

**Nirmeen Zagzoog, MSc, MD**
Neurosurgery Resident
Department of Surgery
Division of Neurosurgery
McMaster University
Hamilton Health General Hospital
Ontario, Canada

**Anna Zicca, MD**
Anaesthesiologist
Department of General Anesthesia and Intensive Care
Anna Meyer Pediatric Hospital
Florence, Italy

# Section I Intracranial Pathology: Tumor

*Hussam Abou-Al-Shaar*

I

# Case 1 Vestibular Schwannoma in Neurofibromatosis Type 2

*Burak Sade and Joung H. Lee*

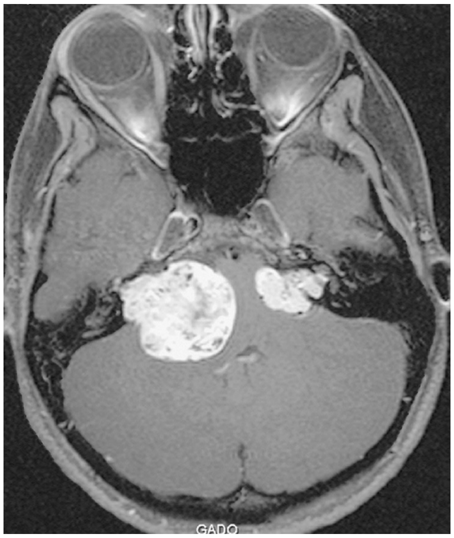

**Fig. 1.1** T1-weighted postcontrast axial magnetic resonance image showing bilateral vestibular schwannomas. Note the severe compression of the brainstem.

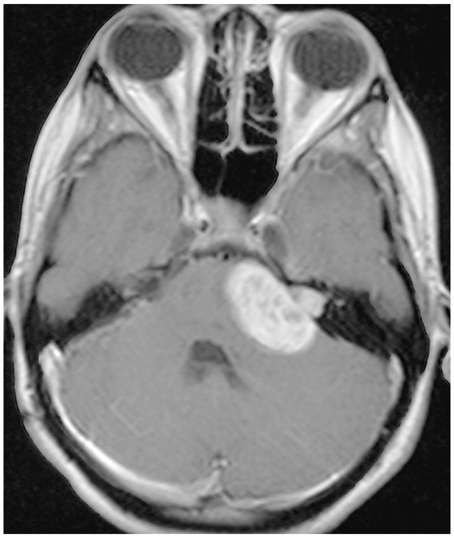

**Fig. 1.2** T1-weighted postcontrast axial magnetic resonance image 2 years after surgery showing complete resection of the tumor on the right. At this point, the left-sided tumor was treated with gamma knife radiosurgery.

## Clinical Presentation

- An 18-year-old left-handed woman with neurofibromatosis type 2 (NF-2) presents with progression of a known vestibular schwannoma (VS).
- Three years ago, she underwent Cyberknife fractionated stereotactic radiosurgery to treat a right-sided VS. Over the last few months, she describes episodes of imbalance and “blackouts” without loss of consciousness.
- Her family history is significant; both her father and sister have NF-2.
- Neurologic evaluation shows bilateral papilledema, decreased hearing on the left side (but still serviceable), and complete loss of hearing on the right side.
- MRI of the brain is shown in ►**Fig. 1.1**.

## Questions

1. What are the diagnostic criteria of NF-2?
2. What additional studies are essential for the decision-making process?
3. What are the main management goals in NF-2 patients?
4. What are the management options?
5. What are the most common surgical approaches?
6. What would be your plan for the right-sided tumor?

She was operated on via a suboccipital retrosigmoid approach, and complete removal of the tumor was achieved with preservation of the facial nerve. During her follow-up, the tumor on the left side showed growth (►**Fig. 1.2**), with worsening of her hearing.

7. What would be your plan for the left-sided tumor?
8. What are the outcomes of radiosurgery for VS?

## Answers

1. *What are the diagnostic criteria of NF-2?*
   - Bilateral VS
   - Family history of NF-2 (first-degree relative) plus unilateral VS
   - Family history of NF-2 plus two of the following:
     - Meningioma, schwannoma (nonvestibular), glioma, neurofibroma, juvenile posterior subcapsular lenticular cataract, or opacity[1]
2. *What additional studies are essential for the decision making process?*[2,3]
   - Audiometric evaluation including pure tone audiometry (PTA), speech reception threshold (SRT), and speech discrimination score (SDS) values
   - "50/50" rule can be used for a cutoff value for serviceable hearing. With PTA showing values < 50 dB and speech discrimination with recognition of > 50%
   - CT scan of the brain to assess bony landmarks and for surgical planning. Also assess the size of the internal auditory canal (IAC) and whether or not there is any dilatation of the canal.
   - Other tests that may be helpful include brainstem auditory evoked responses (BSAER) as preoperative baselines, electronystagmography (ENG), stapedial reflex to assess retrocochlear lesion, and cold caloric testing.[2,3]
3. *What are the main management goals in NF-2 patients?*
   - To preserve serviceable hearing as long as possible
   - To decompress the brainstem from the pressure exerted by the right-sided tumor in this case (▶ **Fig. 1.1**)
   - Early counseling about the genetic implications, as well as the need for training in lip reading and sign language techniques
4. *What are the management options*?
   - Conservative approach with observation is not a good option in this case. This approach is used in cases of small tumor with serviceable hearing. It consists of serial observation with MRI or CT every 6 months for 2 years, then annually.
   - Radiation therapy: external beam radiation or stereotactic radiosurgery (SRS). Gamma knife radiosurgery (a type of SRS) can be used in the following cases:
     - Poor operative candidates because of other medical problems, age, etc.
     - Bilateral VS
     - Tumor progression or tumor residual after surgical resection
     - Usually cases of small tumor and/or tumors with serviceable hearing
   - Surgery is the mainstay of therapy. It is indicated in cases of large- or medium-sized tumors with or without serviceable hearing, in signs of brainstem compression, intractable disequilibrium, severe trigeminal symptoms, and hydrocephalus.
   - Note that patients with bilateral VS (as opposed to unilateral) tend to be younger, with larger tumors, worse preoperative hearing, and have greater chances of losing either cranial nerve VII or VIII functions during surgical excision of the tumor.[3,4]
5. *What are the most common surgical approaches?*
   - Suboccipital or retrosigmoid
   - Translabyrinthine
   - Middle fossa
   - Combined approaches[5]
6. *What would be your plan for the right-sided tumor*?
   - Due to the symptomatic increase in size, failure of previous radiation treatment, and absence of hearing, surgery is indicated on the right-sided tumor.
7. *What would be your plan for the left-sided tumor?*
   - Both surgery and radiosurgery are options in this case. Surgery may have the added benefit of removing any mass effect on the brainstem. However, surgery has a significant risk of hearing loss on the only functioning side. Our preferred course of action would be to treat the tumor with SRS. This is to preserve hearing in the only side where it is still present. She was treated with gamma knife radiosurgery.
8. *What are the outcomes of radiosurgery for VS?*
   - This treatment modality has 81% tumor control rate at 15 years with hearing preservation rates of 73% at 1 year and 48% at 5 years.[6] Preservation of hearing in tumors of this size would be very difficult to achieve with microsurgery.[7] Therefore, surgery would not be recommended on this side as the first option, as long as she has serviceable hearing.

## Suggested Readings

1. National Institutes of Health Consensus Development Conference. Neurofibromatosis. Conference statement. Arch Neurol 1988;45(5):575–578
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7. Mohr G, Sade B, Dufour JJ, Rappaport JM. Preservation of hearing in patients with vestibular schwannoma: degree of meatal filling. J Neurosurg 2005;102:1–5

# Case 2 Subependymal Giant Cell Astrocytomas

*Rihaf Algain, Mohammed Saeed Bafaqeeh, Remi Nader, and Ali Alwadei*

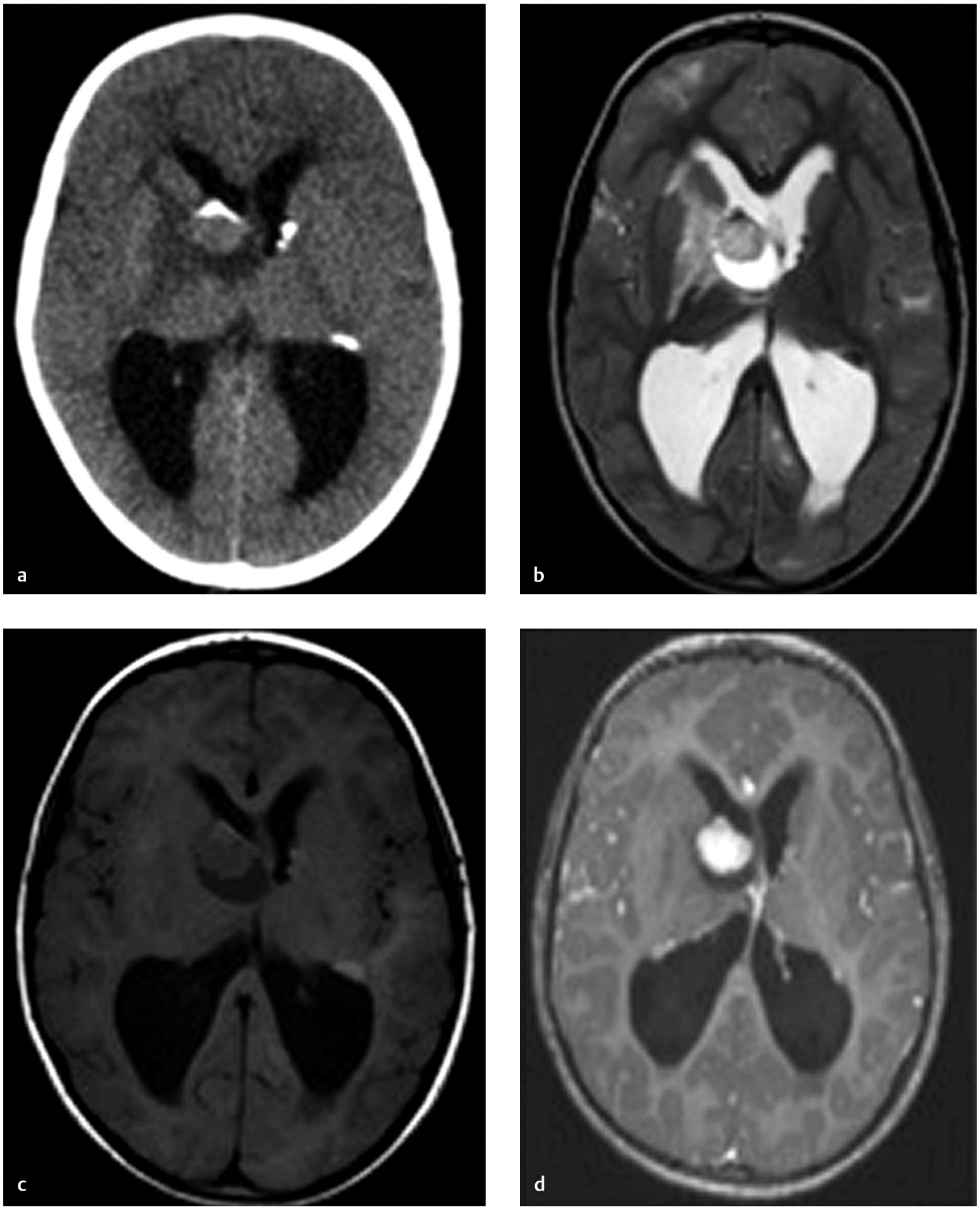

**Fig. 2.1** **(a)** CT and **(b, c, d)** MRI of patient described herein.

## Clinical Presentation

- A 6-year-old boy presents with five days history of headache, vomiting, and increased sleepiness.
- Symptoms started with a generalized tonic-clonic (GTC) seizure, lasting about 2 minutes with a minimal postictal phase.
- On physical examination, he was found to be sleepy but easily arousable, oriented to his parents and place. Cranial nerve examination revealed grade 1 papilledema, with the remaining cranial nerves normal. Power and sensation were normal as well.
- The remainder of the examination is normal.
- Blood work revealed a normal electrolyte profile.
- CT and MRI were performed with pertinent images shown in ▶**Fig. 2.1**.

## Questions

1. Describe the provided images.
2. What is your differential diagnosis? What is the most likely diagnosis and name the syndrome associated with it?
3. What are the diagnostic criteria of tuberous sclerosis (TSC)?
4. Describe briefly the genetics of TSC.
5. Describe the brain lesions that can be associated with TSC and their characteristics.
6. What is your initial management for this patient?
7. What is your management of the intraventricular subependymal giant cell astrocytomas (SEGA)?
8. What are the different surgical approaches to SEGA, what approach would you use?
9. Describe some intraoperative considerations that should be kept in mind to facilitate your surgical management.
10. What is the expected general goal of treatment in a patient with TSC? What are some alternative medical management options to be considered?

## Answers

1. *Describe the provided images.*
    - Axial CT (▶**Fig. 2.1a**) and MRI images (▶**Fig. 2.1b–d**) demonstrate an isointense intraventricular mass near the foramen of Monro with an area of calcification within the anterior portion of the mass. Accompanying hydrocephalus (enlarged ventricular size) and mass effect along the basal nuclei and the septum pellucidum can be appreciated. Calcified subependymal lesions can be seen along the left lateral ventricle.
    - The mass is heterogenous and hypo- to isointense to grey matter on T1-weighted images as well as heterogenous and hyperintense to grey matter on T2-weighted images (▶**Fig. 2.1b, c**); the calcified components is hypointense.
    - There is marked contrast enhancement on T1-weighted images with gadolinium. ▶**Fig. 2.1d**).
2. *What is your differential diagnosis? What is the most likely diagnosis and name the syndrome associated with it?*

    The mnemonic is "CENTRAL MS":
    - C - Choroid plexus papilloma or carcinoma, colloid cyst, central neurocytoma, cavernoma
    - E - Ependymoma, epidermoid/dermoid
    - N - Neurocytoma
    - T - Teratoma, tuber
    - R - "rule out" infection
    - A - Astrocytoma, arteriovenous malformation (AVM), aneurysm, abscess
    - L - Lipoma, lymphoma
    - M - Metastasis, meningioma
    - S - Subependymoma, SEGA

    This is most likely a SEGA that arose from a subependymal nodule of the lateral ventricule as a manifestation of TSC.[1]
3. *What are the diagnostic criteria of TSC?*
    - See ▶**Table 2.1** for criteria
    - The definitive diagnosis is based on the following:
        - Two major criteria
        - One major criterion and two or more minor criteria
    - The possible diagnosis is based on the following:
        - One major criterion
        - Two minor criteria (▶**Table 2.1**)
4. *Describe briefly the genetics of TSC.*
    - TSC is an autosomal dominant disorder.
        - It is caused by mutation in tumor suppressor genes *TSC1* (located on 9q) or *TSC2* (located on 16p)
        - The protein products of these genes are:
            - Hamartin (TSC1)
            - Tuberin (TSC2)
        - Characterized by the formation of benign lesions
        - It involves multiple systems: brain, kidney, liver, skin, heart, and lungs.
        - Incidence: estimated to be 1:6,000.
5. *Describe the brain lesions that can be associated with TSC and their characteristics.*
    - SEGA:
        - Benign tumor (World Health Organization grade I)
        - Glioneural origin
        - Occurs in 10 to 20% of TSC patients
        - Major cause of TSC-related morbidity and mortality

**Table 2.1** Diagnostic criteria of tuberous sclerosis

| Major Features | Minor Features |
|---|---|
| Hypomelanotic macules | "Confetti" skin lesions |
| Angiofibromas or fibrous cephalic plaque | Dental enamel pits |
| Ungual fibromas | Intraoral fibromas |
| Shagreen patch | Retinal achromic patch |
| Multiple retinal hamartomas | Multiple renal cysts |
| Cortical dysplasias | Nonrenal hamartomas |
| Subependymal nodules | |
| Subependymal giant cell astrocytoma | |
| Cardiac rhabdomyoma | |
| Lymphangioleiomyomatosis | |
| Angiomyolipomas | |

*Source:* Adapted from Northup et al 2013.[3]

## Answers (*continued*)

- Cortical tubers:
  - Collection of abnormal neurons and glia
  - Located in the cortex
  - Pathognomonic for TSC
  - Stable throughout life
  - Associated with seizure and autistic spectrum disorder
  - Present in 80 to 100% patients with TSC
  - Infratentorial in 25% cases
- Subependymal nodules (SENs):
  - Intraventricular calcified protrusions (lateral ventricles)
  - Small
  - Asymptomatic
  - Seen in > 90% of patients with TSC

6. *What is your initial management for this patient?*
   - Admission for seizure control and evaluation of hydrocephalus/ventriculomegaly
   - Consider prompt or urgent placement of external ventricular drain until investigations are completed or surgical removal of the mass is performed
   - Start dexamethasone (10 mg IV followed by 4 mg orally Q8 hours) (Galicich)[3,4]
   - Ranitidine 150 mg orally every 12 hours
   - Anticonvulsant and urgent pediatric neurology consultation for management of seizure and epilepsy that can be associated with this syndrome
   - Ophthalmology and endocrinology evaluations
   - Obtain a vascular study such as MRI with MR angiography (MRA), CT scan with CT angiography (CTA)
   - Basic laboratory panel: complete blood count (CBC), electrolytes, prothrombin time (PT), partial thromboplastin time (PTT), type, and screen. Consider obtaining tumor markers: carcinoembryonic antigen (CEA), α-fetoprotein (AFP), beta human chorionic gonadotropin (β-hCG)
   - Other TSC management based on symptoms
7. *What is your management to the intraventricular SEGAs in this patient?*
   - The only known treatment of SEGAs is surgical as they lack responsiveness to other strategies such as chemotherapy or radiation.
   - When complete resection is achieved recurrence is unlikely.[4]
   - The management decision will depend on many factors including the anatomy and the size of the lesion, the patient symptoms, age and the ventricular size, and the surgical team's experience.
   - When SEGA is small and does not seem to be invasive and the patient do not have symptom, it can be followed with serial clinical examination and MRI, as rapid growth (not common) and symptoms can be dealt with on an elective manner; some authors, however, recommend a resection as the surgical morbidity and mortality is low.[5]
   - When SEGA is large and invasive, or when the patient is symptomatic, surgical resection in addition to medical treatment, if required, is performed.
   - This child developed new symptoms related to increased intracranial pressure, which require urgent intervention.
8. *What are the different surgical approaches to SEGA, what approach would you use?*
   - The anatomy and size of the lesion and the child's symptoms, age, and the size of the ventricle are the factors that can be used to decide the surgical approach.
   - The aim is complete removal of the tumor when safe.
   - There are multiple approaches to the lateral ventricle which include: transcallosal, transcortical, and endoscopic approaches. Both the transcallosal and transcortical approaches can be performed via an open conventional craniotomy or by using minimally invasive approaches.

## Answers (*continued*)

- Transcallosal approach: a horseshoe skin incision is made over Kocher's point. A craniotomy of about 6 cm is planned, located two-thirds in front and one-third behind the coronal suture. The dura is reflected with its base toward the sagittal sinus. Major draining veins must be evaluated with vascular imaging preoperatively so as to avoid their injury. Mild adhesion could be encountered as the surgeon dissects along the interhemispheric fissure toward the corpus callosum. The pericallosal arteries and the glistening white surface of the corpus callosum will be appreciated after identifying the cingulated gyrus. The corpus callosum is exposed and incised to a length of 2 to 2.5 cm behind the genu to access the lateral ventricle. The next important step is to evaluate and understand the anatomical landmarks in the ventricular space and their relations to the lesion; the thalamostriate vein and foramen of Monro are used for this purpose. If the vein appears to the right of the foramen, then the right lateral ventricle has been entered; if it appears to the left, then the left lateral ventricle has been entered; and if no vein is visualized, then one should consider the possibility of a cavum septum, as per imaging studies.
- The tumor is resected carefully using microsurgical and microdissection techniques. Care should be taken so as not to injure the underlying structures along the floor of the lateral ventricle. Irrigation is usually used to slow down the venous and tumor bleeding before coagulation.

9. *Describe some intraoperative considerations that should be kept in mind to facilitate your surgical management.*
   - SEGA can invade or be significantly adherent to adjacent structures such as major deep veins, internal cerebral vein, fornix, internal capsule, head of the caudate, and thalamus. Care should be taken not to injure these structures by studying the imaging tests preoperatively and by using a neuronavigation system intraoperatively.
   - Cavitron™ ultrasonic surgical aspirator (CUSA) may facilitate tumor resection by reducing the internal tumor volume, which then creates a more pliable structure that is easily manipulated and dissected from adjacent elements.
   - Careful dural closure with the possible use of previously harvested periosteal graft or a dural substitute is an important step in reducing postoperative complications such as infection and cerebrospinal fluid leakage.
10. *What is the expected general goal of treatment in a patient with TSC? What are some alternative medical management options to be considered?*
    - The goal of treatment for patients with TSC is the maintenance of a good quality of life via both surgical and medical treatment. With regards to medical management, considerations should be given to mTOR inhibitors, such as rapamycin, which has been reported to decrease the volume of the SEGAs and the seizure frequency.

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# Case 3 Sturge–Weber Syndrome

*Faisal Al-Otaibi and Remi Nader*

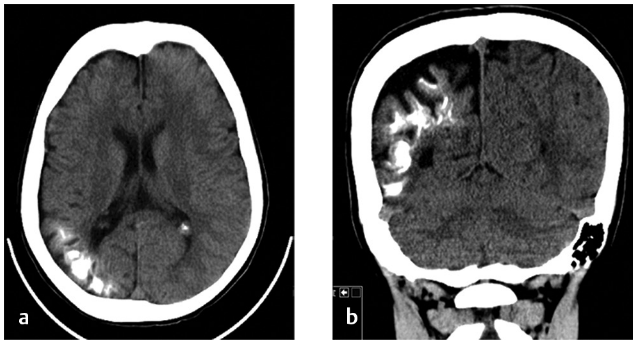

**Fig. 3.1** (**a, b**) Axial and coronal computed tomography without contrast.

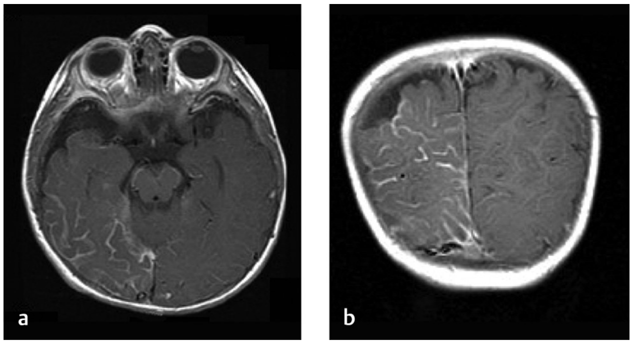

**Fig. 3.2** (**a, b**) Magnetic resonance imaging, axial and coronal T1 with gadolinium, depicting an abnormality in the right hemisphere.

## Clinical Presentation

- A 10-year-old boy presents to the emergency room with a generalized seizure. Upon arrival to the hospital, he was in status epilepticus. He was intubated and received diazepam and phenytoin. His initial CT brain scan was interpreted as “a questionable intracranial hemorrhage.” ▶**Fig. 3.1a, b** shows the abnormality.
- Subsequently, when the family arrived, a further detailed history was obtained. He is known to have a seizure disorder since the age of 4. He has right hemiparesis since his early childhood and was having a poor performance in school . An MRI was obtained and pertinent images are shown above. (▶**Fig. 3.2**).
- The child recovered well next day and was back to his usual neurological status.

## Questions

1. Interpret the initial CT scan.
2. What is the differential diagnosis?
3. What is the most likely diagnosis and the pathogenesis of this condition?
4. What other findings do you expect on general and neurological examination?
5. What is the name of the abnormality shown in ▶**Fig. 3.3a**?
6. What other investigations would you like to obtain?
7. How do you manage status epilepticus?
8. What is the general treatment of this condition?
9. Describe the intraoperative findings shown in ▶**Fig. 3.3b–d**.
10. Describe some surgical options for the treatment of epilepsy in this condition.
11. What are the common indications for hemispherotomy?

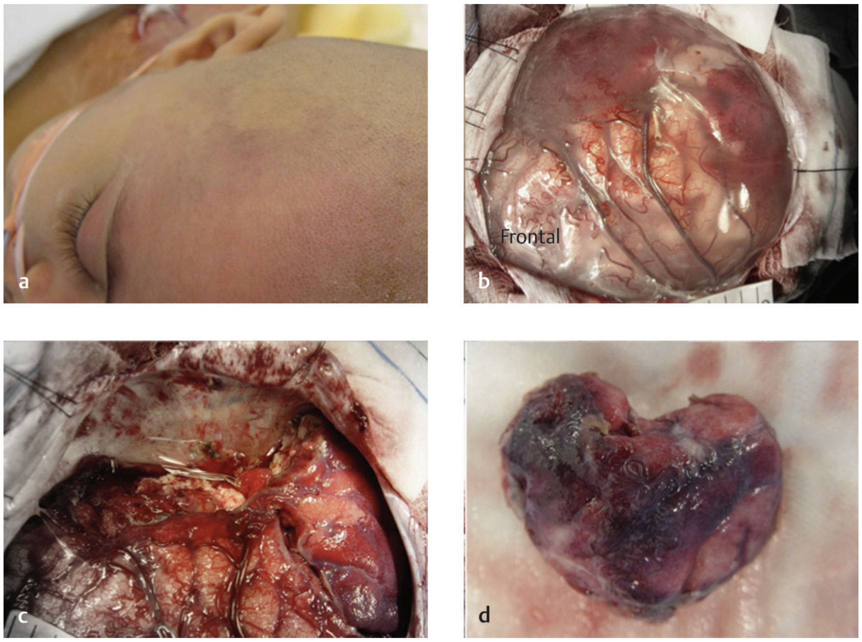

**Fig. 3.3** (**a**) A photograph of the child skin abnormality. (**b**) Intraoperative exposure of the right hemisphere showing the gross cortical abnormality. (**c**) Intraoperative photo showing postsurgical intervention status. (**d**) Intraoperative photo for a specimen from the neocortex depicting the abnormality.

## Answers

1. *Interpret the initial CT scan.*
   - Hyperdensities are seen involving the right hemisphere at the temporal and occipito-parietal regions.
   - These are following the sulci configuration around the gyri (gyriform), and the densities are suggestive of calcifications rather than blood.[1]
   - Overall, this feature is a classical "tram-line" or "tram-track" calcification radiological appearance.
   - There is associated cerebral atrophy.
2. *What is the differential diagnosis?*
   - Congenital: neurocutaneous disorder (Sturge–Weber syndrome)
   - Infectious: encephalitis (viral, bacterial), purulent meningitis
   - Traumatic: contusion, subarachnoid hemorrhage
   - Tumor: gliomatosis, metastatic disease, lymphoma
   - Inflammatory: progressive multifocal leukoencephalopathy, ossifying meningoencephalopathy
   - Vascular: arteriovenous malformation, venous angiomas
3. *What is the most likely diagnosis and the pathogenesis of this condition?*
   Sturge–Weber syndrome. From an embryological prospective, the syndrome has been suggested to result from failure of the primitive cephalic venous plexus to regress in the first trimester.[1]
4. *What other findings do you expect on general and neurological examination?*
   - Port-wine stain (facial angioma): unilateral face involvement at the entire V1 distribution (ophthalmic division of the trigeminal nerve). It is highly predictive of the underlying neurological and ocular disorders and rarely bilateral.[2,3]
   - Extracephalic port-wine stain (52%)[4]
   - Leptomeningeal angiomas: mainly at the parieto-occipital region. The encephalofacial angiomatosis has been classified into three types (Roach scale; ▶**Table 3.1**).[5]
   - Hemiparesis (25–60%) and hemiatrophy (possibly from chronic cerebral hypoxia)
   - Seizures (by the age of 5 years: 95%)[1,4]
   - Mental retardation or learning disability (50–75%)[6]
   - Glaucoma (30–70%) and vascular eye abnormalities
   - Hemianopsia (40–45%)
   - Migraine–like headache (the prevalence in children of < 10 years of age is significantly higher than the general population at 31 versus 5%, respectively)[7]
   - Moyamoya disease
   - Arteriovenous malformation of the lung and liver
   - Endocrinopathy (18-fold increase in the incidence of growth hormone deficiency above the general population)
5. *What is the name of the abnormality shown in* **Fig. 3.3a***?*
   Port-wine stain (facial angioma)
6. *What other investigations would you like to obtain?*
   - MRI/magnetic resonance angiography (MRA) of the brain
   - Electroencephalogram
   - Positron emission tomography (PET) scan or single-photon emission computed tomography (SPECT)
   - Ophthalmology for visual field test[8]
   - Hormonal profile (thyroid function test and growth hormone)
   - Laboratory tests: complete blood count (CBC), electrolytes, glucose level, coagulation profile, and type and screen

**Table 3.1** Roach scale: classification of encephalofacial angiomatosis[5]

| Type | Leptomeningeal Angiomatosis | Facial Angiomatosis | Glaucoma |
|---|---|---|---|
| I | Present | Present | Present/absent |
| II | Absent | Present | Present/absent |
| III | Present | Absent | Usually absent |

## Answers (*continued*)

7. *How do you manage status epilepticus?*
    - Stabilize patient (airway, breathing, and circulation), monitor vital signs, assess oxygenation, and give oxygen through face mask.
    - Initiate electrocardiography.
    - Run laboratory tests: CBC, electrolytes, serum glucose, serum calcium level, and arterial blood gas.
    - Collect finger-stick blood glucose. If glucose < 60 mg/dL, give intravenous (IV) glucose 2 mL/kg D25 water for children > 2 years; for children < 2 years, give 4 mL/kg D12.5 W IV.
    - Perform neurological examination.
    - Pharmacologic treatment (as follows, in the order given):
        - Lorazepam IV (0.1 mg/kg/dose, max: 4 mg/dose, may repeat dose once) or the use of diazepam (0.15–0.2 mg/kg/dose, max: 10 mg/dose)
        - Phenytoin: loading dose of 20 mg/kg
        - Phenobarbital: drip load 15 mg/kg
        - Phenobarbital: drip 15 mg/kg if seizure did not arrest in 30 minutes
        - General anesthesia[9]
8. *What is the general treatment of this condition?*
    - Early aggressive treatment of seizure. Consider surgical option in refractory cases
    - Stroke prevention (aspirin, good hydration)
    - Aggressive management of fever
    - Flu vaccine
    - Glaucoma: annual ophthalmology check
    - Headache: symptomatic treatment
    - Facial angioma: dermatology referral
    - Endocrine: monitor growth and thyroid function
9. *Describe the intraoperative findings shown in* **Fig. 3.3b–d**.
    - The lesion shown in **Fig. 3.3d** represents a Leptomeningeal angioma involving mainly temporal and parieto-occipital regions
    - Post posterior quadrant disconnection procedure is shown in **Fig. 3.3b**.
10. *Describe some surgical options for the treatment of epilepsy in this condition.*
    - Disconnective procedures:
        - Functional hemispherotomy
        - Posterior quadrantectomy
        - Corpus callosotomy
    - Resective procedure: topectomy (localized cortical resection of epileptic focus)
    - Neuromodulation: vagus nerve stimulator placement
11. *What are the common indications for hemispherotomy?*
    - Hemimegalencephaly
    - Sturge–Weber syndrome
    - Rasmussen encephalitis
    - Encephalomalacia (traumatic, ischemia)
    - Porencephaly
    - Cortical dysplasia
    - Migration disorders
    - Hemiplegia–hemiconvulsion–epilepsy syndrome

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# Case 4 Von Hippel–Lindau Disease—Hemangioblastoma

*Sami Obaid, Ramez Malak, and Robert A. Moumdjian*

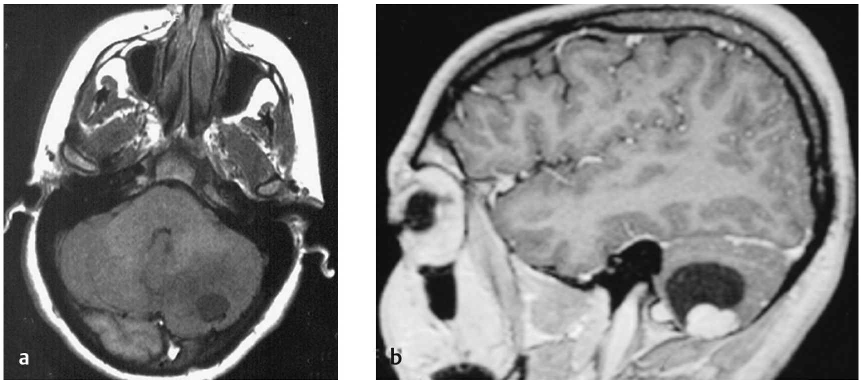

**Fig. 4.1** (**a**) T1-weighted magnetic resonance image (MRI) of the brain, axial cut through the posterior fossa. (**b**) T1-weighted MRI of the brain with contrast enhancement, sagittal cut.

## Clinical Presentation

- A 32-year-old man presents with nausea, vomiting, and headache from the last week.
- He is otherwise neurologically intact.
- MRI scan of his brain is shown in ▶**Fig. 4.1**.

## Questions

1. Interpret the MRI scan shown in ▶**Fig. 4.1**. Name other sequences and/or other imaging investigations that might be helpful in further characterization of the lesion.
2. What is your initial management?
3. Give a differential diagnosis of cerebellar lesions. Which is the most likely?
4. (a) How commonly are hemangioblastomas associated with von Hippel–Lindau (VHL) disease?
   (b) Provide diagnostic criteria of VHL disease.
5. Name other VHL-related lesions, their estimated age of onset, and their frequency.
6. Describe the basic principles of VHL transmission and tumor development. Who should be genetically screened for VHL?
7. Describe the preoperative evaluation for this hemangioblastoma.
8. What are the treatment options?
9. Describe your postoperative follow-up.
10. A spinal MRI demonstrates hemangioblastoma at the T5 level. Describe your management (▶**Fig. 4.2**).

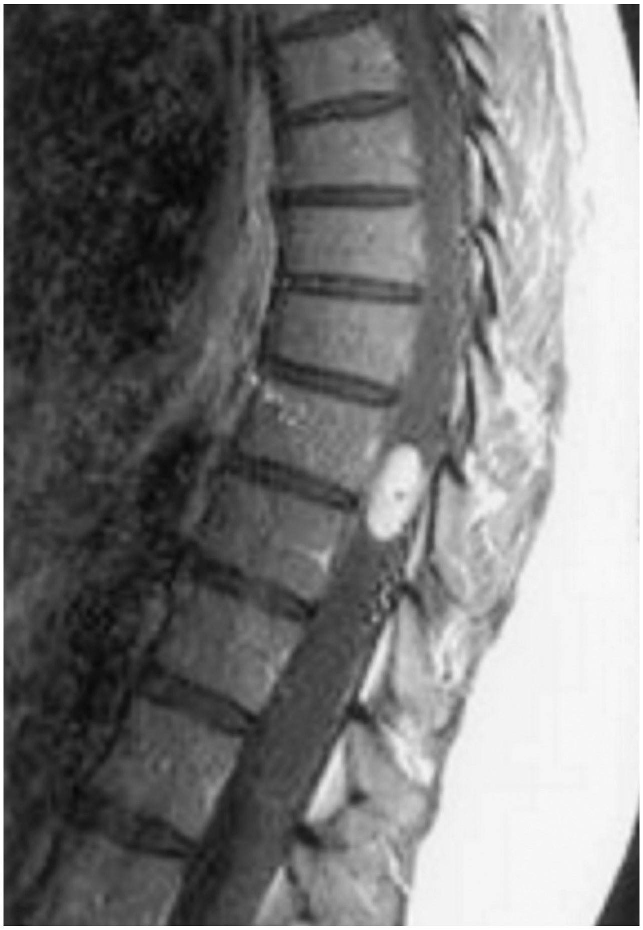

**Fig. 4.2** T1-weighted magnetic resonance image of the thoracic spine with contrast enhancement, midsagittal cut.

## Answers

1. *Interpret the MRI scan shown in ▶* **Fig. 4.1**. *Name other sequences and/or other imaging investigations that might be helpful in further characterization of the lesion.*
   - Nonenhanced T1-weighted MRI scan (▶Fig. 4.1a) shows a cystic lesion in the left cerebellar hemisphere, ~ 1 cm in diameter with limited edema and little mass effect on the fourth ventricle. Although not shown on these images, there was no evidence of hydrocephalus.
   - Sagittal view of a contrast-enhanced T1-weighted MRI scan (▶**Fig. 4.1b**) shows a cystic cerebellar lesion with a densely and uniformly enhancing nodule. The cyst wall does not enhance.
   - Additional sequences include T2-weighted images often revealing flow voids which may help differentiate hemangioblastomas from pilocytic astrocytomas.
   - An important additional investigational study includes vertebral digital subtraction angiography that may show a richly vascularized nodule and feeding vessels, suggestive of hemangioblastoma.
2. *What is your initial management?*
   - Admission and monitoring of the patient.
   - Symptomatic treatment: analgesics, antiemetics, and hydration with IV fluids.
   - Dexamethasone could be indicated in the presence of vasogenic edema.
   - Initial blood workup: complete blood count, electrolytes, coagulation profile, type and screen.
   - Be prepared to perform a ventriculostomy (external ventricular drain or endoscopic third ventriculostomy), if the patient decompensates due to hydrocephalus or increased intracranial pressure.
   - If an urgent shunt is indicated for acute hydrocephalus (rare), keep draining at a low rate (10–15 mL/hour) to avoid overdrainage and upward herniation.
3. *Given a differential diagnosis of cerebellar lesions. Which is the most likely?*
   - Hemangioblastoma (the most likely), metastasis, medulloblastoma, pilocytic astrocytoma
   - Less likely: abscess, glioblastoma, brainstem glioma, cavernous angioma, cerebellar liponeurocytoma, hemorrhage, infarction
   - In the scenario where the tumor extends or originates from the fourth ventricle (not seen in this case), consider the following lesions: ependymoma, subependymoma, and choroid plexus papilloma.
4. *(a) How commonly are hemangioblastomas associated with von Hippel–Lindau (VHL) disease?*
   *(b) Provide diagnostic criteria of VHL disease.*
   - In 25 to 40% of cases, hemangioblastomas are found concomitantly with VHL.[1]
   - Contemporary diagnostic criteria for VHL include both genetic variations and clinical manifestations. The following manifestations are included in the diagnostic criteria:[2–5]
     - Endolymphatic sac tumor
     - Renal cell carcinoma
     - Pheochromocytoma
     - Paraganglioma
     - Glomus tumor
     - Neuroendocrine neoplasm
     - Multiple pancreatic cysts
   - The diagnosis of VHL is established in a patient with:
     - More than two or two central nervous system hemangioblastomas
       OR
     - More than one or one central nervous system hemangioblastoma and one of the clinical manifestations described above
       OR
     - More than one or one of the manifestations described above AND a heterozygous germline pathogenic variant of the VHL gene on chromosome 3p25–26 or a first-degree relative with an established diagnosis of VHL[2–5]
5. *Name other VHL-related lesions, their estimated age of onset, and their frequency.*
   - See ▶**Table 4.1**.[4]
   - Additional tumors have been associated with VHL, including choroid plexus papillomas, pancreatic adenomas and adenocarcinomas, hepatic cysts, and more commonly renal angiomyolipomas.
6. *Describe the basic principles of VHL transmission and tumor development. Who should be genetically screened for VHL?*
   - VHL disease is transmitted in an autosomal dominant fashion with incomplete penetrance. VHL follows the “two-hit” model of hereditary cancer, requiring a second hit (i.e., a hit on both alleles; loss of heterozygosity) in order to lead to tumor formation.
   - Indications for genetic screening for VHL disease remain a matter of debate. Nevertheless, genetic screening should be considered in patients with known VHL-associated lesions (particularly at a young age, i.e., < 30 years) or with first-degree relatives genetically diagnosed with VHL.[2–4]
7. *Describe the preoperative evaluation for the hemangioblastoma.*
   - Craniospinal MRI (cerebellar and spinal hemangioblastomas can be found concomitantly, especially in VHL)

**Table 4.1** Summary of mean age of onset and frequency of CNS and visceral lesions in VHL

| | Mean Age of Onset (years) | Frequency (%) |
|---|---|---|
| **CNS lesions** | | |
| Retinal hemangioblastoma | 25–37 | 15–73 |
| Endolymphatic sac tumor | 22–40 | 3–16 |
| Cerebellar hemangioblastoma | 29–30 | 35–79 |
| Brainstem hemangioblastoma | 25–38 | 4–22 |
| Spinal hemangioblastoma | 33–34 | 7–53 |
| Supratentorial hemangioblastoma | 20–29 | 1–7 |
| **Visceral lesions** | | |
| Renal cell carcinoma | 40–45 | 30–70 |
| Renal cyst | 34–39 | 60 |
| Pheochromocytoma | 20–29 | 16 |
| Pancreatic neuroendocrine tumor | 32–38 | 15–56 |
| Pancreatic cyst | 29–37 | 21–72 |
| Epididymal cyst adenoma | 24 | 25 |

*Abbreviations:* CNS, central nervous system; VHL, Von Hippel–Lindau disease.
*Source:* Data from Chittiboina and Lonser 2015.[4]

## Answers (*continued*)

- Ophthalmologic and audiologic examination
- Abdominal CT scan
- Laboratory work (look for the presence of polycythemia resulting from excessive erythropoietin production by the hemangioblastoma)
- 24-hour urine metanephrine, vanillylmandelic acid, homovanillic acid, and catecholamine test
- Cerebral angiography and embolization (especially for solid highly enhancing nodules) to decrease surgical bleeding[2,3]

8. *What are the treatment options?*
   - Surgery
     - The aim of surgical treatment remains complete removal of the hemangioblastoma.
     - However, multiplicity of lesions and their frequent proximity to vital structures can preclude complete excision. Only symptomatic lesions should be operated while small asymptomatic lesions can be followed with careful surveillance.
     - Surgery is the best option for single accessible lesions. Resection of cerebellar hemangioblastomas leads to symptomatic improvement in up to 98% and absence of recurrence in up to 100% of cases.[6–8]
   - Surgical technique
     - Suboccipital midline incision
     - Suboccipital craniotomy
     - Intraoperative ultrasonography to localize the hypoechoic cyst
     - Y-shaped dural opening
     - Cervicospinal fluid draining via cisterna magna
     - Aspiration of cyst content
     - Extranodular dissection and coagulation of feeding and draining vessels
     - En bloc removal of the nodule
     - The cyst wall should not be necessarily removed as it does not contain tumor
     - Careful closure to avoid cerebrospinal fluid fistulas and to allow easier reopening in cases of recurrences (often associated with VHL)[6]
   - Radiosurgery for hemangioblastomas
     - This should be considered for patients with multiple hemangioblastomas or surgically inaccessible lesions.
     - Different modalities can be used, including stereotactic radiosurgery (SRS), external beam radiotherapy, and proton beam radiotherapy.
     - SRS is generally given in a single dose of 1800 to 2000 cGy. SRS controls the majority of primary and recurrent hemangioblastomas less than 3 cm in size. Response to SRS is enhanced in noncystic hemangioblastomas. Local control is estimated at 82 to 94% at 5 years following treatment.[9,10]
     - However, with cystic tumors, radiosurgery does not reduce the cyst size, and additional surgical removal or repeated evacuations of the cyst may be necessary.[11]
   - Antiangiogenic therapy
     - FGR inhibitors may constitute an option for hemangioblastomas that are not amenable to surgery or radiosurgery.
9. *Describe your postoperative follow-up.*
   - Hemangioblastomas are slow-growing tumors, but a risk of rapidly enlarging cysts is still present and a striking tendency for multiple occurrence (cranial and spinal) is habitual in VHL disease.
   - Pregnancy can be accompanied by the enlargement of a cyst within a few months, sometimes leading to dramatic complications for both mother and fetus.[12]

## Answers (*continued*)

- Regular surveillance includes periodic craniospinal gadolinium-enhanced MRI, abdominal CT scan or ultrasonography, urinary catecholamines, ophthalmologic and audiologic evaluations, for both early diagnosis and follow-up of different manifestations. Rising hematocrit may suggest progression or recurrence. The periodicity of specific examinations depends on age, number, and type of manifestations in each patient. We generally repeat craniospinal MRI every 6 months in the presence of lesions and later yearly if stable.[2,3]

10. *A spinal MRI demonstrates hemangioblastoma at the T5 level. Describe your management (▶* **Fig. 4.2***).*
    - Symptomatic or growing lesions should be treated surgically.[13]
    - Microneurosurgery has improved outcome and safe tumor removal in patients with intramedullary tumors.13
    - Minimally invasive techniques have recently been performed with the advantage of minimizing pain and progressive deformity.[14]
    - Preoperative embolization may be considered.
    - Intraoperative indocyanine green videoangiography can be helpful in identifying feeding vessels intraoperatively.[15]
    - Because of its well-defined margins, careful surgical removal of this benign lesion often provides a cure.[13]
    - Recurrence is rare following total resection, and mild transient neurological deficits are not unusual. The observed deficits often improve within weeks following surgical excision. Overall, symptomatic improvement occurs in two-thirds of patients with sporadic hemangioblastomas.[13]

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# Case 5 Parasagittal Meningioma

*Nirmeen Zagzoog and Almunder Algird*

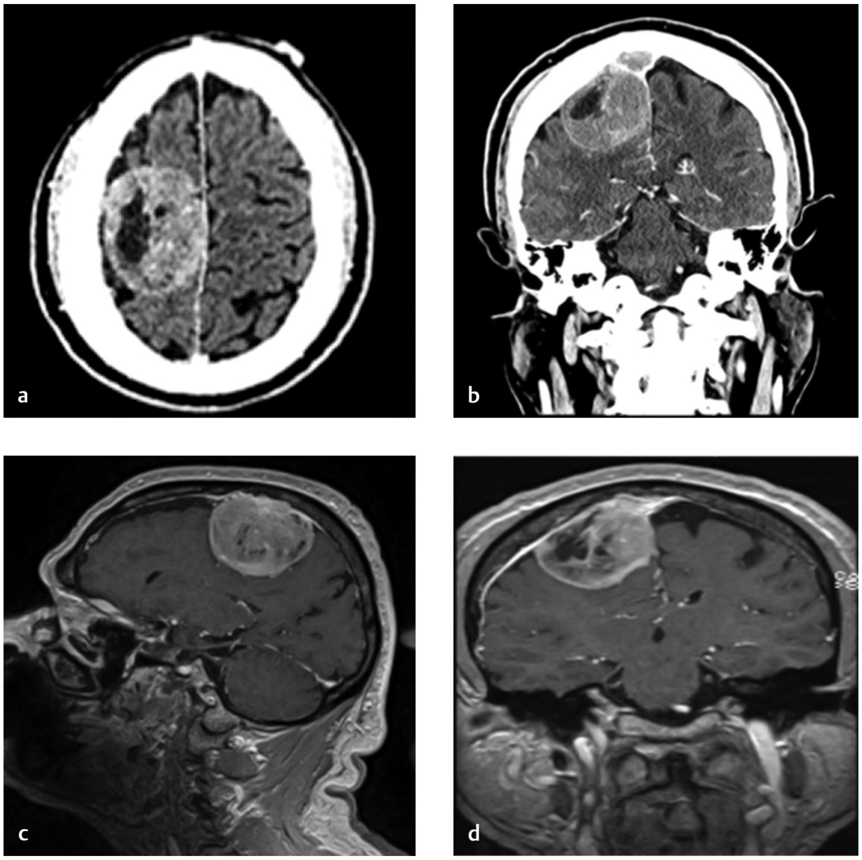

**Fig. 5.1** (**a, b**) Contrast-enhanced computed tomography of the head, and (**c, d**) magnetic resonance images of the brain with gadolinium of patient with left-sided hemiparesis.

## Clinical Presentation

- A 75-year-old right-hand-dominant man presents with gradual onset of left upper and lower extremities weakness for 6 months with rapid deterioration over the past 3 weeks prior to his presentation. He also noticed left paresthesia. His deficits have become severe to the point that he can no longer dress himself. He has difficulty ambulating as a result of the weakness and paresthesia in his left lower extremity.
- His past medical history includes stage I rectal cancer managed surgically with low anterior resection with no subsequent adjuvant therapy 2 years prior to his presentation, diabetes mellitus type II, hypertension, benign prostatic hyperplasia, and colonic polyps.
- On physical examination, speech is normal and cranial nerves are grossly intact. Strength is 4 out of 5 in the major muscle groups of left upper extremity and 3 out of 5 along the left lower extremity. He has full power on the right side. Decreased sensation to light touch and pin prick are noted along the left upper and lower extremities with no specific dermatomal distribution. Reflexes are slightly brisker along the left side as compared to the right. Increased tone is also seen along the left leg more so than the arm.
- A CT head scan was completed which revealed a right frontal abnormality prompting the ordering of a CT head scan with contrast (see ►**Fig. 5.1a, b**).

## Questions

1. Describe the images (▶**Fig. 5.1a, b**).
2. What is your differential diagnosis? What is the most likely diagnosis?
3. What further imaging studies would you request?
4. An MRI with gadolinium is obtained, describe the findings (▶**Fig. 5.1c, d**).
5. What is your management recommendation?
6. What are the indications for surgery?
7. What preoperative measures can be taken to help decrease intraoperative vascular risk/complications?
8. What is the most serious surgical vascular complication specific to this case? How can you avoid it? How do you manage it?
9. How do you grade the surgical resection of meningiomas? What is its clinical implication?
10. You operated and the mass was confirmed to be grade I meningioma. How do you manage postoperative residual tumor? Would your management differ for grade II or III meningioma?
11. Describe a classification system for a meningioma invasion of the superior sagittal sinus.
12. What are the adjuvant treatments of meningioma?
13. What is the outcome of radiosurgery treatment?

## Answers

1. *Describe the images* (▶**Fig. 5.1a, b**).
   A contrast-enhanced CT scan shows a large right-sided parasagittal frontal convexity heterogeneously enhancing extra-axial mass with invasion into the adjacent superior sagittal sinus. Multiple enhancing cystic spaces are seen within the lesion, mostly involving the superolateral aspect of the lesion.
2. *What is your differential diagnosis? What is the most likely diagnosis?*
   - Differential diagnosis includes: meningioma, hemangiopericytoma, lymphoma, pleomorphic xanthoastrocytoma, and metastatic disease.
   - The most likely diagnosis is parasagittal meningioma.
3. *What further imaging studies would you request?*
   - MRI with gadolinium, MR angiography/MR venography (MRV) are used to evaluate the vasculature and examine superior sagittal sinus patency.[1,2]
   - MR spectroscopy (MRS) can be used to examine tumor type, but its usefulness in the diagnosis of meningioma is marginal.[3]
4. *An MRI with gadolinium is obtained, describe the findings* (▶**Fig. 5.1c, d**).
   A large, heterogenous solid and cystic extra-axial enhancing mass approximately 4 × 6 cm in size is observed and located within the right high frontal convexity with invasion into the adjacent superior sagittal sinus.
5. *What is your management recommendation?*
   Treatment options include:
   - Surgical resection since the patient is symptomatic from the lesion
   - Observation with serial MRIs if the patient declines surgery
   - Radiosurgery: this tumor is considered too large for radiosurgery. Surgery is considered a better approach in this case for treatment as the tumor is easily accessible and is located between the first and second thirds of the superior sagittal sinus.

   Surgery is recommended in this case. The patient should be started on steroids (dexamethasone 10 mg IV loading dose, then 4 mg IV/PO every 6 hours) to alleviate the cerebral edema from the brain tumor. Gastric prophylaxis should accompany dexamethasone.

   A discussion with the patient about the risks, benefits, and possible complications about the proposed intervention is necessary.[4,5]

   If the patient consents to surgery, then preoperative anesthesia consult in this case is warranted; in addition to a complete blood workup including coagulation profile.
6. *What are the indications for surgery?*
   - To obtain tissue diagnosis
   - For symptomatic relief from mass effect, and/or to address neurological symptoms and signs due to the tumor
   - Accessible locations
   - Patient medically fit to undergo surgical intervention via craniotomy
   - Increase in size > 1 cm per year or significant radiological changes such as surrounding vasogenic edema with mass effect, and midline shift[4]
7. *What preoperative measures can be taken to help decrease intraoperative vascular risk/complications?*
   - Preoperative angiography (CTA, MRA, DSA) to evaluate the arterial supply should be performed and it can be combined with tumor embolization to reduce intraoperative blood loss and decrease surgical time.[6,7]
   - Placing a precordial ultrasound probe prior to the start of the surgery can help in identifying an air embolus early intraoperatively especially in high-risk cases.
   - Reviewing the vascular anatomy on the MRI and the surface vein relationships to the actual tumor and adjacent cortical structures and gyri is also essential in planning the surgery.

## Answers (*continued*)

- Using neuronavigation and combining the vascular and other imaging studies can help in adequate planning of the opening and approach.

8. *What is the most serious surgical vascular complications specific to this case? How can you avoid it? How do you manage it?*

   Intraoperative venous sinus injury can cause significant hemorrhage, air embolism, or venous infarction.

   Avoidance is achieved by:

   - Obtaining detailed preoperative venous imaging (MRV or venogram) to assess patency or degree of sinus invasion by the tumor
   - Using neuronavigation to better define sinus location
   - Being very careful during opening not to injure the sinus while drilling/cutting the bone or raising the bone flap, as it might be adherent to the dura in some cases
   - Avoiding extensive retraction on the sinus or large draining veins during tumor resection and being careful not to cause sinus occlusion

   Management of venous sinus injury is first by preparation and having a strategy to repair the sinus. Ask anesthesia to monitor $PCO_2$ for air embolism so you can treat a potential air embolus. When sinus injury occurs, control bleeding by packing with pledgets of hemostatic material, have temporalis fascia or muscle graft ready to use if packing fails or vascularized pericranium, and use precordial Doppler probe. Vascular clamps and aneurysm clips can be used as a last resort because of their risk of injury to sinus walls and afferent (bridging) veins.[5,8]

9. *How do you grade the surgical resection of meningiomas? What is its clinical implication?*

   The Simpson grading system can be used to grade the extent of surgical resection as follows:

   - Grade I: macroscopically complete removal with excision of dural attachment and abnormal bone (including sinus resection when involved)
   - Grade II: macroscopically complete resection with endothermy coagulation (cautery or laser) of dural attachment
   - Grade III: macroscopically complete tumor resection without resection or coagulation of dural attachment or of its extradural extensions (e.g., hyperostotic bone)
   - Grade IV: partial removal leaving tumor in situ
   - Grade V: simple decompression (± biopsy)

   The extent of surgical resection is the most important prognostic factor for tumor recurrence. Five-year recurrence rates are as follows: 10% for Simpson grade I, 20% for grade II, 30% for grade III, and 40% for grade IV.

   Overall, histological type, size of tumor, and Simpson grade are significant independent prognostic factors for recurrence.[9,10]

10. *You operated and the mass was confirmed to be grade I meningioma. How do you manage postoperative residual tumor? Would your management differ for grade II or III meningioma?*
    - Grade I (pathology): observation and follow-up with serial MRI with consideration of surgical resection if significant increase in size occurs.
    - Grade II: closer postoperative follow-up with serial MRI at more frequent intervals. In cases of recurrence or progression of residual tumor, reoperation or radiosurgery is recommended.
    - Grade III: radiosurgery is typically recommended.[11,12]
11. *Describe a classification system for a meningioma invasion of the superior sagittal sinus.*
    - Type I: lesion attachment to the outer layer of the sinus wall
    - Type II: tumor fragment inside the lateral recess
    - Type III: invasion of the ipsilateral wall
    - Type IV: invasion of the lateral wall and roof
    - Types V and VI: complete sinus occlusion, without or with invasion of the contralateral wall, respectively[5]
12. *What are the adjuvant treatments of meningioma?*
    - Radiation therapy
    - It is indicated for residual or recurrent tumors, malignant pathologies, and small-volume tumors in critical location. It can arrest tumor growth.[12,13]
    - Chemotherapy
      - It has modest efficacy; therefore, it is confined to recurrences that are refractory to radiotherapy and are considered inoperable.
      - Options include: antiprogesterone (mifepristone = RU-486), hydroxyurea, interferon-α 2B, and Sandostatin long-acting release.[14]
13. *What is the outcome of radiosurgery treatment?*
    - Five-year local control rate of 93% for benign, compared with 68 and 0% for patients with atypical or malignant meningiomas, respectively.
    - Treatment-related complications include: cranial nerve deficits, symptomatic parenchymal changes, internal carotid artery stenosis, symptomatic cyst formation, and decrease in functional status.
    - Tumor volume, tumor margin dose, or previous radiotherapy is not associated with the development of radiation-related complications.[15]

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# Case 6 Tuberculum Sellae Meningioma

*Mazda K. Turel, Naif M. Alotaibi, and Fred Gentili*

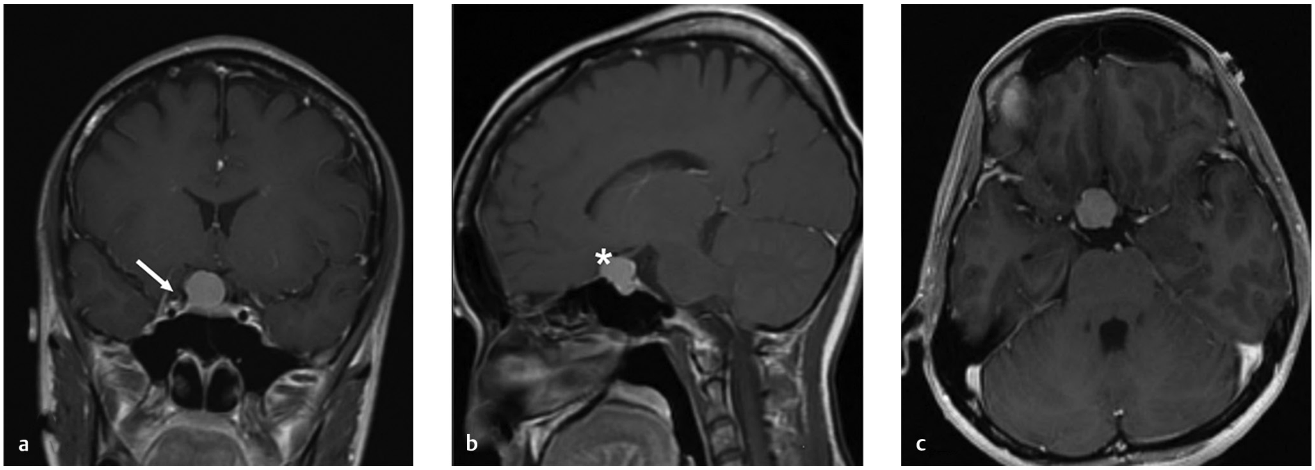

**Fig. 6.1** (**a**) Coronal, (**b**) sagittal, and (**c**) axial gadolinium-enhanced MRI of a 46-year-old female showing a 2-cm tuberculum sellae meningioma with no involvement of the cavernous sinus (*arrow*), displacement of the optic nerve superiorly (*asterisk*) without invasion into the optic canal, and no encasement of the anterior cerebral artery making it an ideal case for endoscopic endonasal excision.

## Clinical Presentation

- A 46-year-old woman has a history of progressive painless diminution of vision in the right eye for approximately 1 year.
- Formal visual examination revealed a visual acuity of 20/200 in the right eye and 20/20 in the left eye with a right inferior temporal field deficit.
- Fundoscopic examination revealed primary optic atrophy in the right eye.
- The rest of the neurological examination was unremarkable.

## Questions

1. Describe the pertinent findings on the MRI.
2. What are the most common modes of presentation of this tumor?
3. What are the options for the management of this tumor?
4. What are the indications for surgery?
5. Is primary radiosurgery an option in the treatment of this tumor?
6. What is the goal of surgery?
7. Enumerate the preoperative investigations specific to this pathology in preparing this patient for surgery.
8. What are the different surgical approaches that could be used to remove this tumor?
9. What are the advantages and disadvantages of the endonasal endoscopic approach?
10. Briefly discuss the important technical nuances in removal of this tumor endoscopically.
11. What are the possible intraoperative and postoperative complications?
12. Discuss briefly the radiological and functional (visual and endocrine) outcomes after endoscopic excision of tuberculum sella meningiomas.

## Answers

1. *Describe the pertinent findings on the MRI* (▶**Fig. 6.1**).
   The MRI shows an enhancing dural-based lesion arising from the tuberculum sellae. It is hypointense on T1 and hyperintense on T2-weighted images with bright contrast enhancement. It arises in the midline slightly eccentric to the right where it displaces the right optic nerve superiorly. There is no significant lateral extension nor does it encase the right internal carotid artery or the anterior cerebral artery superiorly. There is no extension into the right optic canal or cavernous sinus involvement. There is no evidence of brain edema surrounding the tumor. The MRI findings are in keeping with a diagnosis of tuberculum sellae meningioma.
2. *What are the most common modes of presentation of this tumor?*
   Progressive painless visual loss is the most common mode of presentation. This may be unilateral or bilateral with either a homonymous or bitemporal hemianopsia. Patients with larger-sized tumors may also have endocrinological dysfunction either on the basis of compression of the pituitary stalk or the gland itself. Less common modes of presentation include persistent headache or seizures.

   Clinically, these tumors may be differentiated from olfactory groove meningiomas since tuberculum sellae meningiomas initially present with visual dysfunction and as they grow larger, they can affect olfaction; whereas olfactory groove meningiomas usually present with disturbances in olfaction and as they enlarge posteriorly, they may affect vision.
3. *What are the options for the management of this tumor?*
   - Observation for a smaller tumor found incidentally with no visual disturbance
   - Surgery
   - Radiation therapy
4. *What are the indications for surgery?*
   - Progressive diminution of vision is the main indication
   - Persistent headache
   - Mass effect
   - Surgery is rarely done for seizures alone
   - Endocrine dysfunction alone is uncommon and not commonly an indication for surgery
5. *Is primary radiosurgery an option in the treatment of this tumor?*
   Due to the intimate proximity of the tumor to the optic nerves and chiasm, which usually overlie it, and the fact that visual loss is a common presentation, radiosurgery is usually not considered as a primary line of treatment for this tumor. If there were extension into the sella, radiosurgery to the gland could lead to delayed hypopituitarism.

   However, after adequate tumor removal and optic nerve decompression, if there is residual tumor either within the cavernous sinus or adherent to the internal carotid artery, radiosurgery could be considered as an option, especially if the pathology denotes a higher-grade tumor or demonstrates growth in follow-up imaging.

   In patients with severe comorbidities where surgery can carry a very high operative risk, fractionated radiosurgery may be considered as the primary option.
6. *What is the goal of surgery?*
   Safe gross total resection with preservation of all neurovascular structures and reversal of visual and endocrinological dysfunction is the goal of surgery.

   However, this is not always possible and the goals need to be individualized for each patient depending on the age, nature of presenting symptoms, size, and radiological features such as the extensions of the tumor, vessel encasement, intraoperative findings, and the availability of adjuvant treatments.
7. *Enumerate the preoperative investigations specific to this pathology in preparing this patient for surgery.*
   - A detailed history and physical examination followed by an ophthalmological evaluation with visual acuity, visual field testing, and fundoscopic examination to look for primary optic atrophy. An ENT evaluation may be obtained if the endoscopic approach is being considered.
   - A full pituitary hormonal and biochemical profile to detect any preoperative endocrine dysfunction, which may need to be optimized before surgery
   - MRI of the brain with special focus on the sella and suprasellar region to delineate tumor extension, especially its lateral extension, encasement of vessels, displacement of the pituitary stalk, and presence of brain edema
   - If an endonasal approach is being considered, a CT with thin slice axial and coronal reconstruction to evaluate extent of hyperostosis and study endonasal anatomy is very helpful.
   - Anesthesia evaluation to ascertain fitness for surgery should also be carried out.
8. *What are the different approaches that could be used to remove this tumor?*
   - Transcranial open approaches
     - Bilateral (usually reserved for larger-sized tumors > 4 cm)
       - Bilateral subfrontal approach
       - Bilateral transbasal interhemispheric +/- lamina terminalis
     - Unilateral
       - Unilateral subfrontal approach
       - Unilateral subfrontal + orbital osteotomy
       - Pterional transsylvian approach +/- orbitozygomatic
       - Supraorbital mini-craniotomy (keyhole)

## Answers (*continued*)

- Transsphenoidal (TSS)
  - Microsurgical TSS
  - Pure expanded endoscopic
- The diverse growth patterns of this tumor may require that a variety of approaches be considered to provide the optimal surgical management.

9. *What are the advantages and disadvantages of the endonasal endoscopic approach?*
   - Advantages
     - It has more direct approach to midline central skull base pathology.
     - Via a transcribiform, transplanum, transtubercular, and transclival corridors, it allows access to the entire central anterior skull base.
     - Minimal or no brain retraction is required.
     - Provides early en-route devascularization of the tumor.
     - In contrast, transcranial procedures often require working over and around the optic nerves and chiasm. With the endoscopic approach, there is no manipulation of these structures.
     - It allows for visualization and careful dissection of the superior hypophyseal and anterior cerebral arteries.
     - It provides a panoramic view of relevant anatomy with improved illumination and a better cosmetic result.
   - Disadvantages
     - It is a technically challenging approach with a steep learning curve.
     - There is restricted maneuverability and workspace making instrument manipulation difficult.
     - There is a lack of binocular vision during this approach.
     - Cerebrospinal fluid (CSF) leak remains a significant challenge requiring complex reconstruction techniques.
10. *Briefly discuss the important technical nuances in removal of this tumor endoscopically (*▶**Fig. 6.2**).

    The following are important points and nuances that should be considered when utilizing the endoscopic approach:
    - The initial harvesting of a vascularized nasoseptal flap, which is a major element in the reconstruction[1]
    - Wide exposure of both lateral and medial optic-carotid recesses, the parasellar carotids, optic prominences, planum, sella, and clivus
    - Devascularization of the dura with coagulation of the posterior ethmoidal arteries, which commonly feed the tumor
    - Use of a bimanual microsurgical technique, internal debulking of the tumor, and careful extracapsular dissection
    - Initial dissection of the anterior superior margin followed by the lateral margins

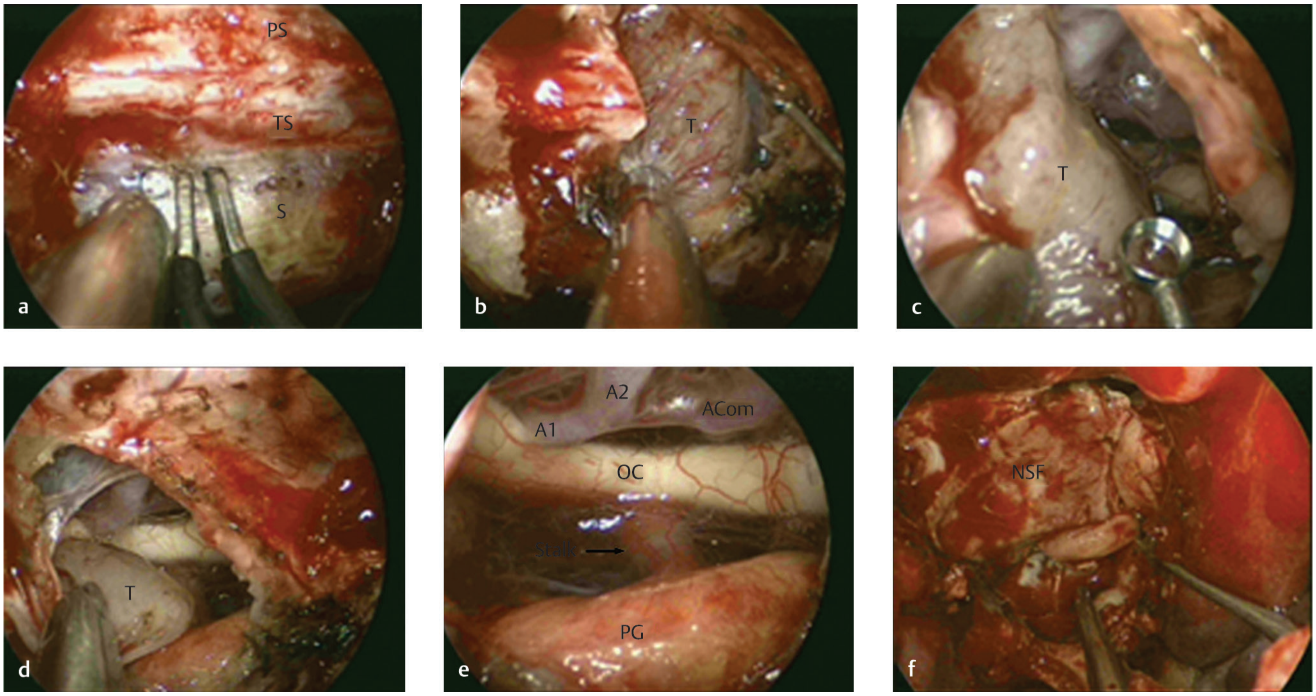

**Fig. 6.2** Intraoperative images showing (**a**) the exposure of the sella (S), tuberculum sellae (TS), and the planum sphenoidale (PS). (**b**) The tumor (T) is visible after opening the dura. It appears pale due to elimination of its blood supply during the exposure. (**c, d**) Extracapsular dissection of the tumor off the optic nerve and anterior cerebral artery after internal debulking. (**e**) Excellent view of the A1, A2, and ACom, optic chiasm (OC), pituitary stalk, and pituitary gland (PG) after complete tumor removal. (**f**) Reconstruction of the defect with the vascularized nasoseptal flap (NSF).

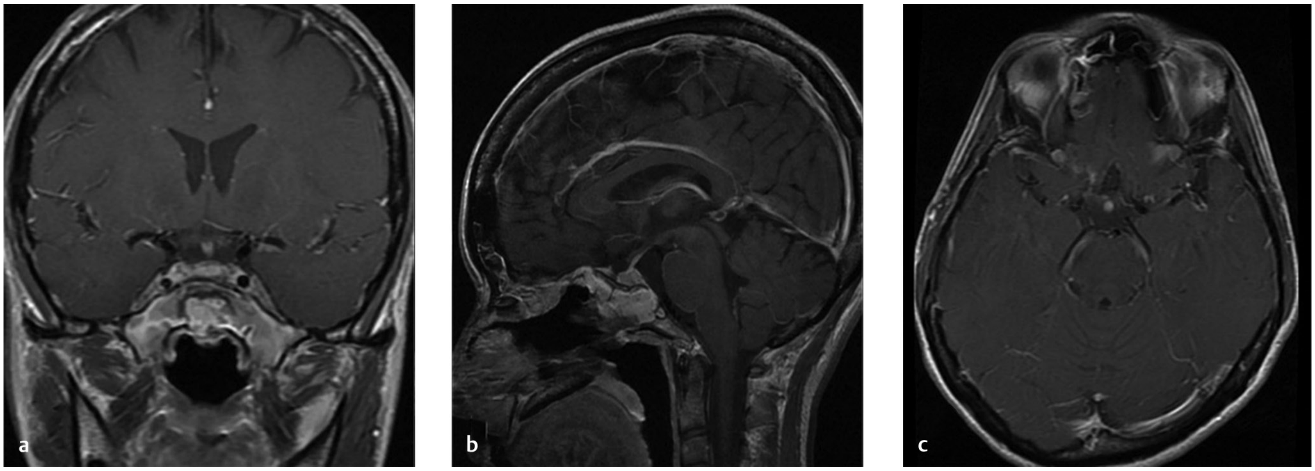

**Fig. 6.3** (**a**) Coronal, (**b**) sagittal, and (**c**) axial gadolinium-enhanced postoperative MRI showing no residual tumor.

## Answers (*continued*)

- Visualization of key neurovascular structures including the optic nerves, chiasm, anterior cerebral arteries, and anterior communicating complex
- Dissection of the inferior margins where the pituitary stalk is usually visualized either in the midline or displaced to one side
- If there is evidence of optic canal invasion on the preoperative MRI, the optic sheath is opened medially taking care to avoid damage to the ophthalmic artery[2]
- The tumor is then delivered into the sphenoid sinus and removal completed.
- A 30-degree scope can be used for inspection of any residual tumor.
- Meticulous hemostasis is achieved after copious irrigation.
- A multilayered reconstruction concludes the procedure. A postoperative MRI scan shown in ▶**Fig. 6.3** exemplifies adequate resection vi the above approach and techniques.

11. *What are the potential intraoperative and postoperative complications?*
    - Intraoperative
        - Vascular damage to the hypophyseal arteries or perforators from the A1 portion of the anterior cerebral artery can cause significant morbidity.
        - Attempts to remove tumor adherent to the internal carotid artery may cause a vascular injury.
        - Manipulation of the pituitary stalk can result in transient diabetes insipidus.
        - Inadvertent manipulation of the optic nerve or its blood supply can cause visual deterioration, which most often is transient but could be permanent as well.
    - Postoperative
        - CSF leak is the most common complication, but its incidence has significantly declined with improved reconstruction techniques including a multilayered closure including the use of the vascularized nasoseptal flap.
        - Meningitis, which can occur as a result of CSF leak.
        - Pneumocephalus secondary to a CSF leak, while uncommon, may result in altered level of consciousness.
        - Electrolyte and hormonal disturbance need to be checked for and corrected in the immediate postoperative period.
12. *Discuss briefly the radiological and functional (visual and endocrine) outcomes after endoscopic excision of tuberculum sella meningiomas.*
    - Among the case reports and small case series published in literature in the last decade (about 200 patients), the gross total resection rates vary from 54 to 100% with an average of 81%. CSF leak rates are 15 to 20%. Endocrinological complications include transient (2.6%) or permanent diabetes insipidus (1.9%) and postoperative hypopituitarism (3.3%).[3]
    - In a meta-analysis comparing the open versus the expanded endonasal approaches for tuberculum sellae meningiomas, visual improvement was much higher (73.5 vs. 58.7%) in the endoscopic cohort. CSF leak rates were higher in the former group as well (21.3 vs. 4.3%)[4]
    - The endoscopic approach yields higher visual improvement and lower visual deterioration rates.
    - Although postoperative CSF leak remains a significant problem, the incidence has substantially reduced with the routine use of the vascularized pedicled nasoseptal flaps.
    - The removal of intradural skull base meningiomas remains controversial and a clear understanding of the limitations of the endoscopic approach is very important and helps guide patient selection for the use of this technique.

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# Case 7 Olfactory Groove Meningioma

*Lissa Marie Peeling, Stephano Chang, and Stephen J. Hentschel*

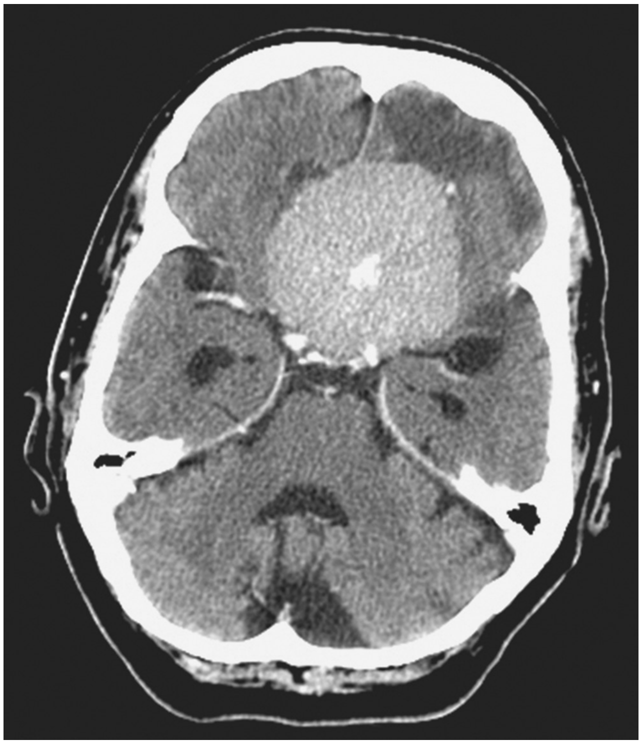

**Fig. 7.1** Axial CT scan of the brain with contrast.

## Clinical Presentation

- A 78-year-old woman presents with increasing difficulties with ambulation and memory deficits. The patient also complains of progressive visual loss for an unspecified period.
- Medical history is significant for atrial fibrillation and hypothyroidism treated with Eltroxin (GlaxoSmithKline, Brentford, London, UK), digoxin, and warfarin.
- On examination, the patient was confused and agitated. She had a right temporal visual field cut and decreased acuity in the left eye. Her examination was otherwise unremarkable but difficult to assess because of her confusion.
- A CT scan of the head was performed (▶**Fig. 7.1**). The patient was then admitted for further management.

## Questions

1. What is your differential diagnosis?
2. What is your initial management?
3. What is Foster–Kennedy syndrome?
4. What further imaging studies, if any, would you request?
5. MRI scan is obtained; please describe the findings (▶**Fig. 7.2**).
6. What is the main feature differentiating an olfactory groove meningioma and a tuberculum sellae meningioma?
7. Describe the pros and cons of the most common operative approaches for olfactory groove meningiomas.
8. Describe the general operative technique *via craniotomy* including management for the arterial supply and plan for resection.
9. What is the expected prognosis of this lesion following your treatment? What intraoperative steps can you take to minimize recurrence of the lesion?

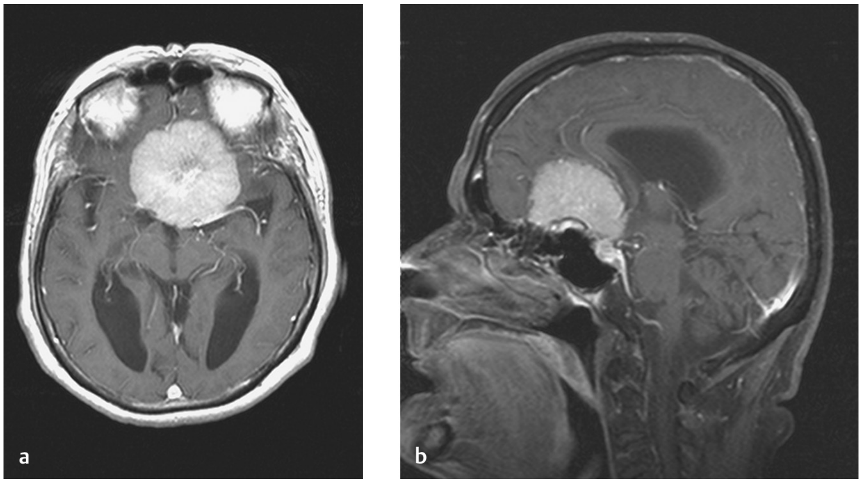

**Fig. 7.2** (**a**) Axial T1-weighted MRI plus gadolinium of the brain. (**b**) Sagittal T1-weighted MRI plus gadolinium of the brain.

## Answers

1. *What is your differential diagnosis?*
   - Skull-base meningiomas (olfactory groove meningioma, tuberculum sellae meningioma, and planum sphenoidale meningioma)
   - Pituitary adenoma
   - Craniopharyngioma
   - Olfactory groove metastases, schwannoma, or hemangiopericytoma
2. *What is your initial management?*
   - Reversal of warfarin in preparation for operation with vitamin K and fresh frozen plasma or prothrombin complex concentrate
   - Corticosteroid: dexamethasone—loading dose of 10 mg intravenously (IV) followed by 4 mg IV/PO (by mouth) every 6 hours
   - Preoperative assessment for fitness of surgery
3. *What is Foster–Kennedy syndrome?*
   - Foster–Kennedy syndrome has been described with olfactory groove meningiomas, although a minority present with all of its features.[1,2] The components of this syndrome include:
     - Anosmia
     - Unilateral optic atrophy
     - Contralateral papilledema
4. *What further imaging studies, if any, would you request?*
   - Further imaging studies should include an MRI scan with MR angiography.
   - There is no need for conventional catheter angiography because the relationship of the vessels of the lesion should be well defined in noninvasive angiography.
5. *MRI scan is obtained; please describe the findings (▶* **Fig. 7.2***).*
   - The T1-weighted MRI sequence with gadolinium demonstrates a large diffusely enhancing tumor arising from the anterior skull base with the anterior cerebral arteries located posterior to the lesion and the optic chiasm located inferior to the lesion.
6. *What is the main feature differentiating an olfactory groove meningioma and a tuberculum sellae meningioma?*
   - The main feature distinguishing olfactory groove meningiomas and tuberculum sellae meningiomas is the location of the chiasm. The optic nerves and chiasm are located inferolateral to the tumor in olfactory groove meningiomas but are located superolateral to the tumor in tuberculum sellae meningiomas.[1]
7. *Describe the pros and cons of the most common operative approaches for olfactory groove meningiomas.*
   - Subfrontal approach ± orbital osteotomies
     - For larger tumors (> 3 cm), a bicoronal flap is turned.
     - For smaller tumors (< 3 cm), a unicoronal flap is turned.[1]
     - Advantages:
       - Early devascularization along the skull base with division of feeding vessels[3]
       - Allows for access into orbits to coagulate the ethmoidal arteries that supply the majority of the tumor[4]
       - Orbital osteotomies minimize frontal lobe retraction[1]
       - Allows for harvesting of vascularized pericranium for skull base reconstruction
       - May provide greatest likelihood of Simpson I/II resection when performed with orbital osteotomies[2]
     - Disadvantages:
       - Opens frontal sinus, increasing the risk of postoperative cerebrospinal fluid (CSF) leak and infection[3]
       - Sacrifice of anterosuperior sagittal sinus
   - Pterional
     - Advantages:
       - Early exposure of optic apparatus and carotid artery prior to tumor manipulation[1,3,5]
       - Early access to basal cisterns for CSF drainage for brain relaxation[4]

## Answers (*continued*)

        - Shorter distance to tumor[5]
        - Avoids entry into frontal sinus[1]
        - Spares venous structures[3,5]
        - Less frontal lobe retraction unless orbital osteotomies are performed with subfrontal approach[1]
    - Disadvantages:
        - Narrow working angle[5]
        - May be blinded in upper portion of tumor, which may require extensive frontal lobe retraction[5]
        - Difficult to access ethmoid arteries[3]
        - Difficult to repair basal skull defects[3]
- Interhemispheric
    - Advantages:
        - Preserves superior sagittal sinus
        - Frontal sinus not opened
        - Careful dissection can minimize frontal lobe elevation/retraction
    - Disadvantages:
        - Higher risk of contusion to frontal lobes
        - Operative route is long and narrow
        - Risk to bridging veins
        - Difficult to access vascular supply
- Endoscopic endonasal transcribriform
    - Advantages:
        - Direct exposure of tumor with minimal brain retraction[6]
        - Early devascularization of tumor[6]
        - Easy access to tumor that has invaded paranasal sinuses[6]
    - Disadvantages:
        - Learning curve associated with endoscopic technique[7]
        - Unable to access large intracranial extensions of tumor
        - May have increased rates of CSF leak and recurrence rates compared to open surgical techniques[7]

8. *Describe the general operative technique via craniotomy including management for the arterial supply and plan for resection.*
    - Craniotomy ± orbital osteotomies
    - Early interruption of the blood supply
        - If using a subfrontal approach, isolate and cauterize the anterior and posterior ethmoidal arteries within the orbit to reduce the risk of intraoperative hemorrhage.
    - Gently retract the frontal lobes with exposure of the tumor.[1,3]
    - The tumor capsule must be dissected, cauterized, and opened. The tumor is then debulked using an ultrasonic aspirator.
    - At the posterior aspect of the tumor, an intact arachnoid plane should be identified separating the tumor from the anterior cerebral arteries, chiasm, and optic nerves.
    - Excellent visualization of the anterior cranial fossa floor to permit tumor resection and repair of defects.[1]
9. *What is the expected prognosis of this lesion following your treatment? What intraoperative steps can you take to minimize recurrence of the lesion?*
    - Prognosis
        - Clinically, this patient has a high likelihood of returning to normal mental status with a reversal of her visual changes.[1,2,8]
        - Although recent studies show low recurrence rates following complete resection, these tumors may have a high predilection for late recurrence at the cranial base and sinuses, with rates as high as 23 to 41% at 10 years.[9–11]
        - Due to the increased difficulty and risks associated with reoperations, aggressive primary resection including drilling of hyperostotic bone, removal of dura as well as resection of sinus extension to reduce the chance of recurrence should be a goal of this surgery.[1–10]
        - In these circumstances, reconstruction of the skull base is a necessity to prevent postoperative CSF leaks and meningitis.

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# Case 8 Sphenoid Wing Meningioma

*Abdulmajeed Alahmari, Mahmoud AlYamany, Mohammed Saeed Bafaqeeh, Remi Nader, and Ehtesham Ghani*

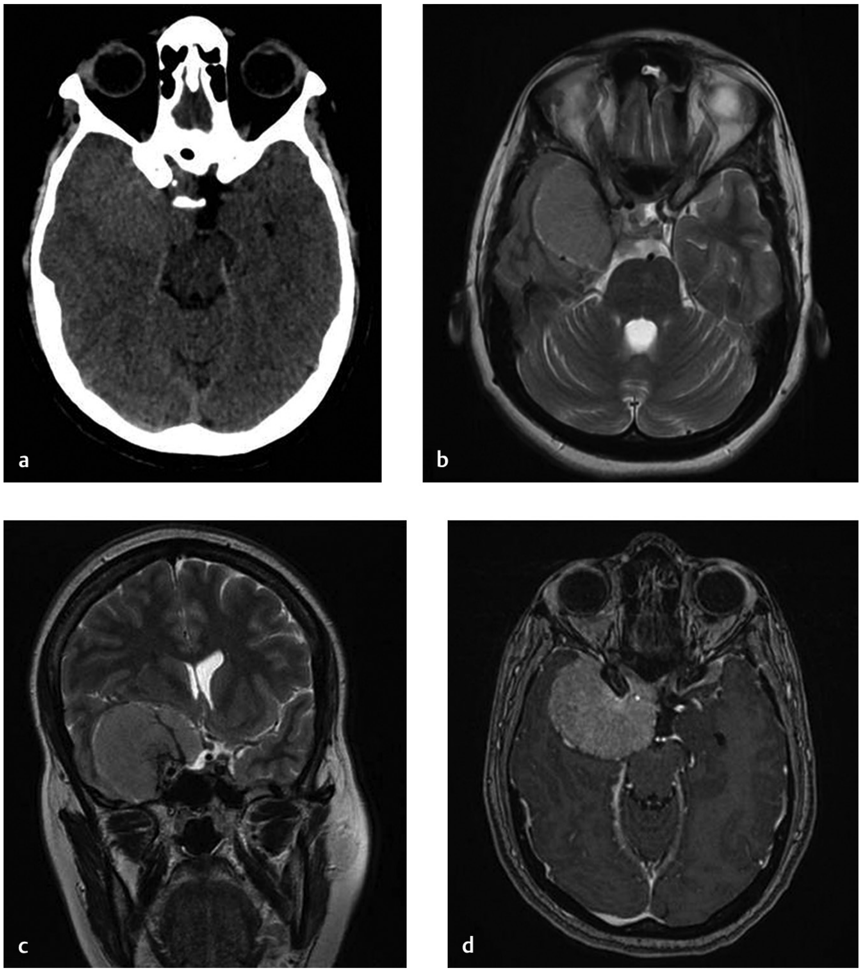

**Fig. 8.1** (**a**) CT and (**b, c, d**) MRI of patient described herein.

## Clinical Presentation

- A 45-year-old right-handed female (mother of four children) presents to the emergency department with first time generalized tonic colonic seizures.
- She reported a 5-months history of decreased vision in the right eye progressively worsening over past 2 weeks, double vision, and sharp pain over the right eye.
- On physical examination, she was found to have visual acuity of “counting fingers at one-meter distance” on the right side, right sixth cranial nerve palsy, and numbness in the distribution of the ophthalmic branch of the right trigeminal nerve.
- The rest of the general and neurological examination was unremarkable.
- Laboratory testing revealed elevated blood glucose level of 22 mmol/L, and low hemoglobin of 8.5 g/dL.
- The patient underwent a CT scan followed by an MRI of the brain (▶**Fig. 8.1a–d**).

## Questions

1. State the findings on the CT and the MRI scans.
2. What is your differential diagnosis?
3. How are these lesions classified?
4. Does this lesion explain the patient symptoms? Please elaborate.
5. What further evaluations (imaging or laboratory studies) specific to this pathology would you request?
6. What is your initial management for this patient?
7. What is the potential arterial blood supply to this lesion?
8. What is your surgical approach to this lesion? List intraoperative steps that can be taken to facilitate your surgical management?
9. After the lesion is completely removed, you have a large dural defect. How do you address this defect?
10. List the factors that may increase the risk of recurrence of this lesion.

## Answers

1. *State the findings on the CT and the MRI scans.*
   - Axial CT (▶**Fig. 8.1a**) and multiplane MRI (▶**Fig. 8.1b–d**) demonstrate a large right sphenoid wing lesion, which is extra-axial, well circumscribed, and has a broad dural base. It extends into the optic canal, interpeduncular cistern, encasing and stretching the internal carotid artery (ICA) as well as the M1 segment of the middle cerebral artery (MCA).
   - It is isodense to the brain on CT with features of hyperostosis involving the sphenoid wing and anterior clinoid process which also can be seen on the CT as well as the MRI images.
   - On MRI, the lesion is isointense to grey matter on T2-weighted images with intense and homogeneous enhancement on contrasted T1-weighted images. Flow void signals can be seen inside the lesion. The gadolinium enhancement in the area of hyperostosis may be related to meningiomatous bone infiltration.
   - Additionally, there is another small lesion along the left sphenoid wing which represents another meningioma.
2. *What is your differential diagnosis?*
   - Primary tumor: sphenoid wing meningioma, hemangiopericytoma, primary bony lesion, chordoma, chondrosarcoma
   - Metastatic or malignant lesion: extra-axial metastases, lymphoma, cranial osteosarcoma, nasopharynx lesion with extension to the skull base
   - Less likely diagnosis include:
     - Infection: brain abscess, subdural empyema
     - Inflammatory condition: sarcoidosis
     - Traumatic: subdural or epidural hematoma
3. *How are these lesions classified?*
   There are many classifications of sphenoid meningiomas, one of which is Cushing's classification which is as follows:
   - Lateral (outer) or pterional
     - Greater chance of en plaque meningioma
     - Higher association with headaches
     - Can be large globular tumors
     - Arterial supply from: superficial temporal artery, middle meningeal arteries, and anterior meningeal artery
   - Middle
     - May become large before they are diagnosed or turn symptomatic
     - Greater association with seizures
     - Arterial supply from branches of ethmoidal arteries
   - Medial or clinoidal
     - Cranial nerves II and III may be affected early
     - Surgically challenging with lower chance of complete resection
     - Arterial supply from ascending branch of pharyngeal artery, recurrent branch of ophthalmic artery via superior orbital fissure
4. *Does this lesion explain the patient symptoms? Please elaborate.*
   - This is a sphenoid wing meningioma that arises from the lesser wing of the sphenoid bone. These lesions can grow in any direction and cause neurological deficit based on the adjacent structures that they compress or encase. The tumor can spread in any of the following directions:
     - Medially compressing or invading the cavernous sinus, compressing the ICA and cranial nerves III, IV, V, and VI causing double vision
     - Superiorly the ICA and its branches outside the cavernous sinus can be encased causing transient ischemic attack (TIA) or stroke
     - Anteriorly through the optic canal or/and above it to the anterior fossa leading to reduced visual acuity and/or causing frontal lobe compression with potential cognitive impairment
     - Laterally compressing the temporal lobe causing seizure and memory impairment
     - Posteriorly compressing the oculomotor nerve and the midbrain in the interpeduncular cistern causing double vision (▶**Fig. 8.2**)
   - Sphenoid bone hyperostosis may also be observed.
5. *What further evaluations (imaging or laboratory studies) specific to this pathology would you request?*
   - MR angiography (MRA) or CT angiography (CTA) to understand the vascular anatomy, in particular the ICA and its branches in relation to the tumor

## Answers (*continued*)

- Cerebral angiography which may include embolization of the feeding arteries and/or balloon occlusion test, if indicated
- Endocrine testing is important as pituitary insufficiency has been reported to occur in 22% of patients with anterior skull base meningiomas
- Ophthalmological evaluation
- Neuropsychology assessment

6. *What is your initial management for this patient?*
   - Prompt admission for potential surgical intervention
   - Start dexamethasone (10 mg IV followed by 4 mg orally Q8 hours)
   - Ranitidine 150 mg orally every 12 hours
   - Anticonvulsant (phenytoin 300 mg orally HS)
   - Ophthalmology examination
   - Endocrinology evaluation and management of her abnormal blood glucose level
   - MRI with MRA, CT scan, and digital subtraction angiography (DSA; 6 vessels angiography)
   - Neuropsychological assessment
   - Preoperative blood testing (complete blood count [CBC], electrolytes, coagulation profile, type and cross match 2 units)
   - Anesthesia evaluation
7. *What is the potential arterial blood supply to this lesion?*
   - The tumor may be vascularized by branches of the external carotid artery (ECA) mainly the middle meningeal artery, the ICA such as meningohypophyseal trunk or both.
   - Although CTA or MRA can be done to evaluate for this propose, our recommendation is to perform a selective catheter angiography to better understand the shape of the ICA, vascularity of the tumor and determine the possibility of preoperative embolization.
8. *What is your surgical approach to this lesion? List intraoperative steps that can be taken to facilitate your surgical management.*
   - Adequate surgical exposure with minimal morbidity is a challenge for skull base lesions.
   - The aim is complete removal of the tumor, and excision of its dural origin and involved bone.
   - The most common approach to remove sphenoid meningioma is a pterional craniotomy.
   - One should also consider extending the craniotomy and performing cranio-orbito-zygomatic approach to better reach the tumor and minimize brain retraction.
   - A generous bone flap is important to allow good exposure for safe and complete removal of the tumor and excision of the involved dura.
   - Drilling of the pterion will facilitate early control of the tumor's main blood supply through dural and pterional branches of the ECA.
   - Drilling of the lateral sphenoid wing up to the superior orbital fissure is usually performed to remove infiltrated bone.
   - Extradural or intradural clinoidectomy and unroofing of the optic canal can be performed to decompress the optic nerve.
   - The dural opening can be performed laterally in the frontotemporal region.
   - Because this tumor is encasing the ICA and M1 segment of the MCA, the M2 or M3 segments should be identified first distally in the upper border at the Sylvian fissure, as it is difficult and risky to try to identify the ICA and its branches proximally early on. A true invasion of the wall of the MCA is very rare and separation of the MCA from the tumor can be achieved in many cases.
   - After good exposure "de-dressing" and "devascularization" of the tumor, one can start internal debulking followed by a careful dissection of the tumor borders.
   - Care should be taken not to injure the adjacent arteries and nerves as well as the major veins such as deep cerebral veins.
   - Cavitron ultrasonic surgical aspirator (CUSA) is quite helpful in resecting the lesion in a piecemeal fashion.
   - Our recommendation is to leave a sleeve of tumor on the ICA, if it is not easily dissectible. The tumor portion that is truly invading the cavernous sinus may be left for later radiation therapy if not easily resectable.
   - If the dura cannot be resected, it should be coagulated; but care should be taken not to coagulate the dura over the falciform ligament (roof of the optic nerve) or the dura that is covering the upper portion of the superior orbital fissure (to prevent third and fourth cranial nerve palsies).
   - Continuous monitoring of blood loss is essential.
   - Neuronavigation is an important safety measure as well as the neurophysiological monitoring.
   - Doppler ultrasound can be used to identify the ICA and MCA before they are exposed during the tumor debulking.
   - Careful dural closure with the possible use of periosteum or artificial dura should be planned.
9. *After the lesion is completely removed, you have a large dural defect. How do you address this defect?*
   - The defect needs to be closed; several options are available for this.
     – Bovine pericardium
     – Fascia lata (requires preparing a harvesting site)
     – Pericranium (also requires harvesting and potentially expanding the incision)
     – Synthetic dural substitute
   - Closure of the defect may be further optimized by using fibrin glue around the suture line to prevent cerebrospinal fluid (CSF) seepage.[10]

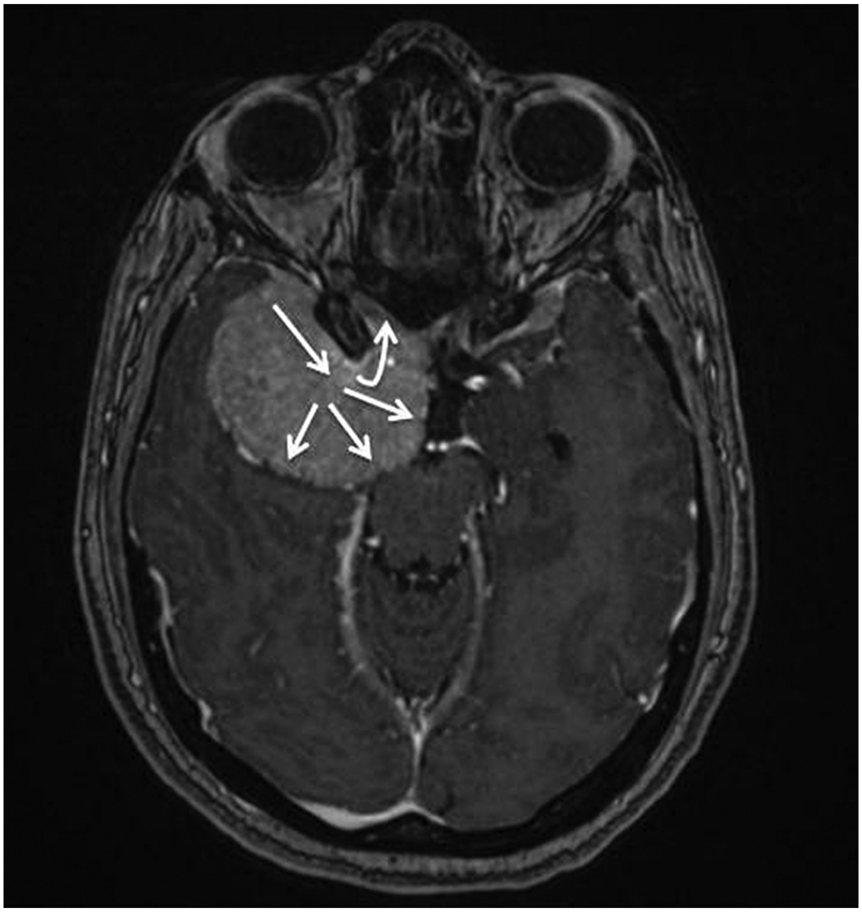

**Fig. 8.2** T1 enhancing image of the same patient. The small left meningioma represents the typical origin of sphenoid wing meningioma (the lesser wing of the sphenoid bone). The large lesion along the right side represents the possible extensions of sphenoid wing meningioma as it might grow (*arrows*): medial extension to the cavernous sinus, sella or suprasellar areas, anterior extension to the optic canal or the anterior cranial fossa, inferior extension to the superior orbital fissure, and posterior extension involving the interpeduncular cistern and the midbrain.

## Answers (*continued*)

10. *List the factors that may increase the risk of recurrence of this lesion.*
    - Extent of resection (Simpson grading)
    - Histological grading:
      - Benign (grade I) with a recurrence rate of 6.9%
      - Atypical (grade II) with a recurrence rate of 34.6%
      - Malignant (grade III) with a recurrence rate of 72.7%
    - Radiation-induced meningiomas
    - Syndromic meningioma (e.g., NF1)
    - Location (medial sphenoid meningioma)
    - Age and gender
    - Pathological marker of high proliferativelesion

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# Case 9 Hemangiopericytoma

*Burak Sade and Joung H. Lee*

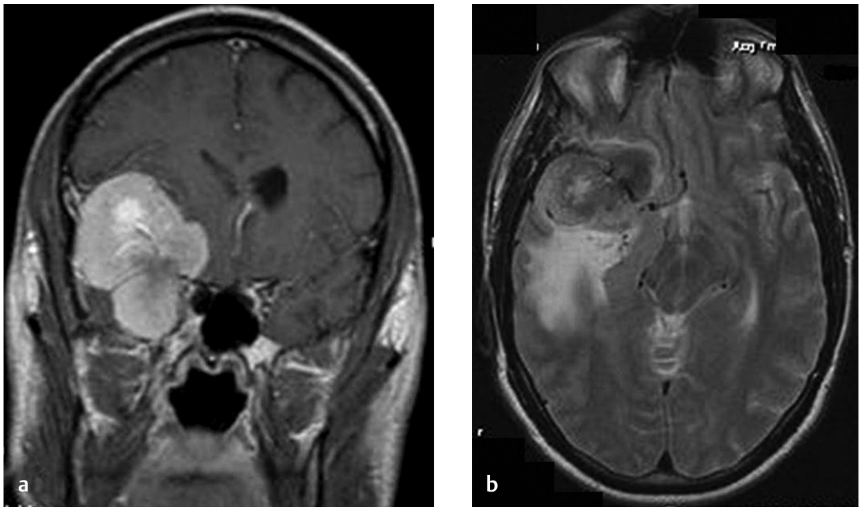

**Fig. 9.1** (**a**) T1-weighted postcontrast coronal MRI showing a diffusely enhancing mass within the right Sylvian fissure. (**b**) Note the presence and caliber of vessels in and around the tumor on T2-weighted images suggestive of hypervascularity.

## Clinical Presentation

- A 42-year-old right-handed male presents with a few months history of headache, memory problems, and decline in cognitive functions.
- His neurological evaluation is significant for short-term memory loss and left homonymous superior quadranopsia.
- MRI of the brain is shown in ▶**Fig. 9.1**.

## Questions

1. What are the significant findings on the MRI?
2. What is your differential diagnosis?
3. Would you consider any other imaging study and if yes then why?
4. What would be your initial management?
5. How is hemangiopericytoma classified in the most recent 2007 WHO classification and how does it differ from meningioma in this classification?
6. Apart from the follow-up of the neurological status of the patient, is there any particular aspect of the postoperative care in this patient that you would like to be cautious about?
7. Is there any role of adjuvant treatment?
8. What is the prognosis of hemangiopericytoma?

## Answers

1. *What are the significant findings on the MRI?*
   Tumor size and location (site of origin), severe edema, midline shift, enhancement pattern, and hypointense areas within the tumor on T2-weighted images suggesting hypervascularity.
2. *What is your differential diagnosis?*
   - Meningioma and hemangiopericytoma
   - Less likely: metastases, glioma, lymphoma, brain abscess
3. *Would you consider any other imaging study and if yes then why?*
   Digital subtraction angiography (DSA) in order to outline the pattern of vascularization (▶**Fig. 9.2**), and to assess the feasibility of preoperative embolization in a highly vascular tumor such as the presented case.
4. *What would be your initial management?*
   - Admit the patient to the ward or intensive care unit.
   - Obtain laboratory studies (complete blood count [CBC], electrolytes, prothrombin time [PT], partial thromboplastin time [PTT], type and screen or crossmatch 4 units of packed red blood cells [PRCB]) if surgery is anticipated.

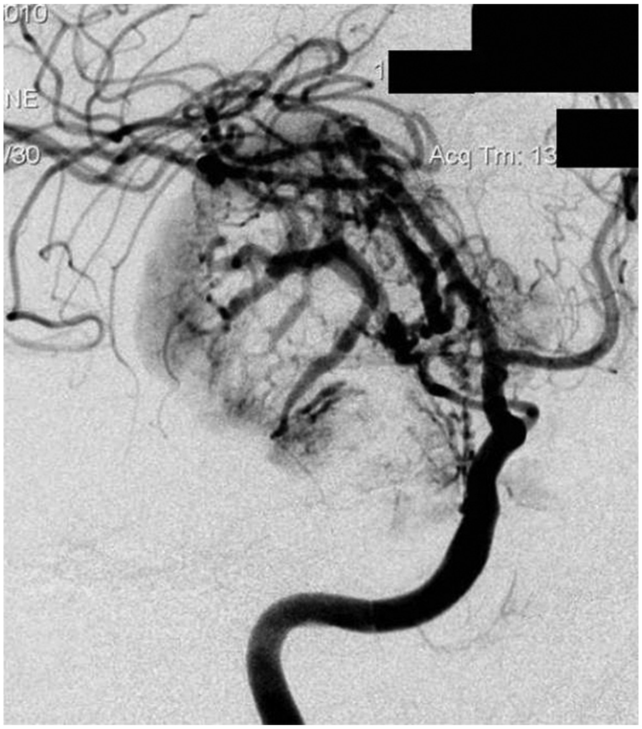

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**Fig. 9.2** Angiogram of the right internal carotid artery demonstrating the presence of significant caliber high-flow tumor feeders from the middle cerebral artery.

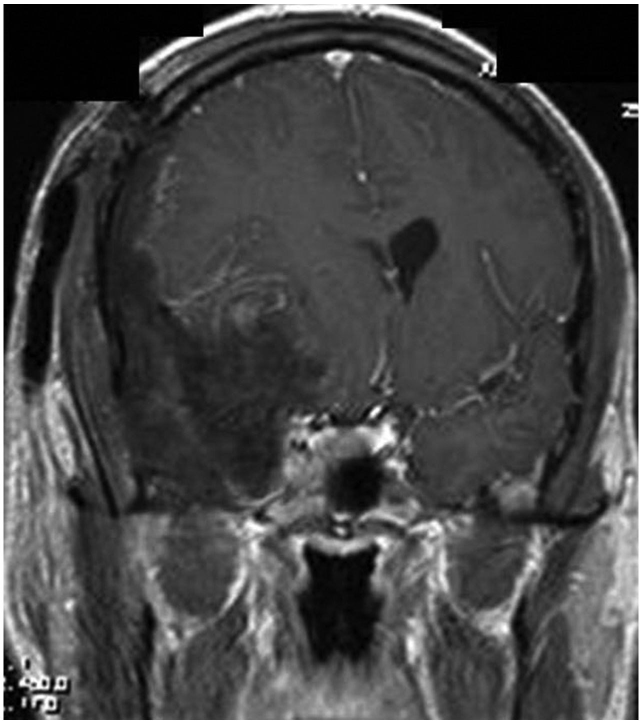
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**Fig. 9.3** Postoperative T1-weighted postcontrast coronal image following gross total resection.

## Answers (*continued*)

- Steroids (dexamethasone). Note though that the impact may be subtle due the multifactorial nature of the raised intracranial pressure in this case (huge tumor size, hydrocephalus, edema).
- Seizure prophylaxis
- Preoperative angiography with possible embolization
- Obtain further imaging to plan surgery, if necessary: CT scan to determine bony landmarks and magnetic resonance angiography (MRA) if unable to get an angiogram.
- Schedule surgery on a semiurgent basis—it may need to be a staged procedure. (Make available sufficient amounts of blood products such as PRCBs, fresh frozen plasma, and packed platelets in anticipation of significant intraoperative blood loss.)
  - The patient was operated through a right frontotemporal craniotomy. The tumor was very hard, rubber-like, and highly vascular necessitating transfusion of significant amount of blood products. Gross total resection was achieved (▶**Fig. 9.3**).
  - The tumor histology confirmed the diagnosis of hemangiopericytoma.

5. *How is hemangiopericytoma classified in the most recent 2007 WHO classification and how does it differ from meningioma in this classification?*
   Both tumors are listed under the main group of “meningeal tumors” as distinct entities. Hemangiopericytoma is defined as a highly cellular and vascularized mesenchymal tumor with a high tendency to recur and metastasize outside the central nervous system.[1] They are either WHO grade II or grade III. Other tumors included in this group are hemangioblastoma, melanocytic lesions, and mesenchymal nonmeningothelial tumors.
6. *Apart from the follow-up of the neurological status of the patient, is there any particular aspect of the postoperative care in this patient that you would be like to be cautious about?*
   As the patient received a significant amount of blood products intraoperatively, one would need to continue monitoring the coagulation parameters as massive transfusions can result in coagulopathy in the postoperative period and therefore necessitate ongoing replacement of the required products.

## Answers (*continued*)

7. *Is there any role of adjuvant treatment?*
   - The role and efficacy of radiotherapy is controversial. Although it has been shown to delay recurrence and improve progression-free survival, no proven effect of radiotherapy has been shown on the overall survival in the literature except for one study.[2–4] In this context, the extent of resection has been shown to have a more significant impact on overall survival.[3,4]
   - There has been no proven role of chemotherapy.
   - Following surgery, the patient received radiotherapy and has been recurrence free for 2 years.

8. *What is the prognosis of hemangiopericytoma?*
   In the series of Rutkowski and colleagues, median survival was 16 years and 1-, 5-, and 10-year survival rates were reported as 100, 92, and 68%, respectively.[3] Median time to recurrence was 5 years with 1-, 5-, and 10-year progression-free survival rates being 96, 49, and 28%, respectively. Incidence of extracranial metastasis was 20%. In the series of Schiariti and colleagues, recurrence was seen in 72% cases with recurrence rates of 3.5% at 1-year, 46% at 5-year, and 92% at 15-year follow-up.[4] Extraneural metastasis was reported as 26%. Spine, long bones, liver, lung, and abdominal cavity are the most common sites of metastasis.

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# Case 10 Anterior Clinoidal Meningioma

*Imad N. Kanaan*

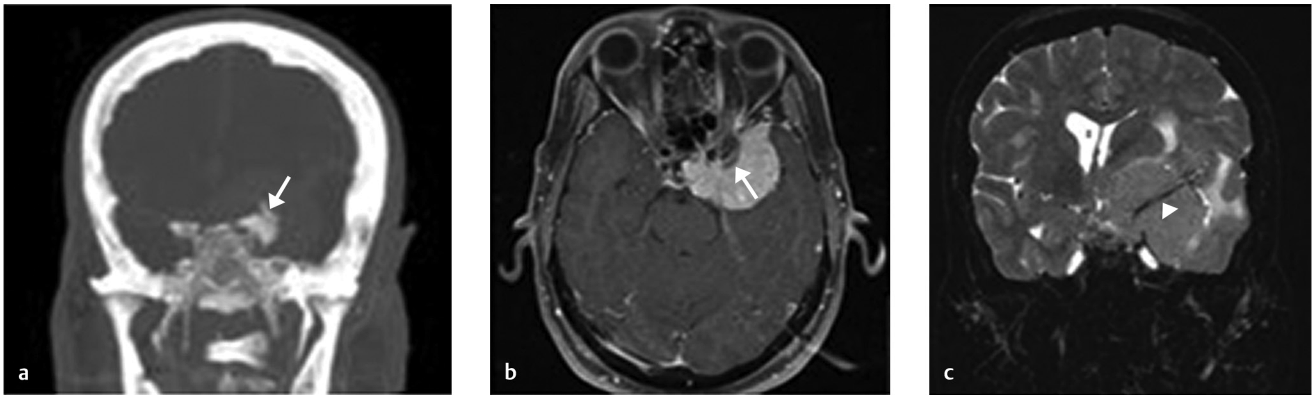

**Fig. 10.1** (**a**) CT scan coronal, (**b**) MRI axial view with gadolinium, (**c**) MRI coronal T2, depicting anterior clinoid hyperostosis (*white arrow*) associated with the mass lesion encasing the internal carotid artery (*arrow head*).

## ■ Clinical Presentation

- A 67-year-old female, known to have several systemic comorbidities including a history of right breast cancer resection followed by chemotherapy and hormonal therapy, was diagnosed a year later with follicular thyroid carcinoma and underwent a hemithyroidectomy. The patient has been in remission for the past 2 years.
- She presented recently to the emergency room with a 5-day history of headache, dizziness, progressive left-sided weakness/paresis with strength of 2/5 (Medical Research Council [MRC] grading), and right-sided partial ptosis.
- An ophthalmology evaluation revealed moderate decrease in visual acuity bilaterally: OD: 20/50 and OS: 20/40.
- CT scan and MRI of the brain were ordered (▶**Fig. 10.1**).

## ■ Questions

1. What is your diagnosis and how would you grade this lesion?
2. How do you explain the clinical findings in this case, based on the radiographic and anatomical observations?
3. How do you determine the site of origin for larger meningiomas in these regions?
4. What are your management goals, treatment options, and future follow-up plans?
5. What are the important investigations and medical management measures prior to surgery that are specific in the case of this lesion?
6. What are the generally recommended steps in the neurosurgical treatment of these lesions?
7. How would you handle bleeding from the cavernous sinus intraoperatively?
8. What are the general outcomes after removal of this tumor?
9. What are some postsurgical considerations when faced with an inability to totally remove the tumor?

Management

- The patient underwent a tailored fronto-orbito-zygomatic (FOZ) craniotomy and partial resection of an anterior clinoidal meningioma (ACM) with decompression of her right optic nerve.
- Age, comorbidities, and severe encasement of the cerebral vessels hampered our ability to achieve the intended excision of the meningioma, resulting in a subtotal resection. Postoperatively, the patient reported no major improvement in her neurological finding. The pathology report was meningothelial meningioma.

**Table 10.1** Anterior clinoidal meningioma classification[2,3]

| Al-Mefty Group | | Suprasellar Extension | | |
|---|---|---|---|---|
| Group I | –Lower clinoid<br>–Arachnoid plane (−) | A<br>≤ 2cm | B<br>2–4cm | C<br>≥ 4cm (giant) |
| Group II | –Lateral clinoid<br>–Arachnoid plane (+) | | | |
| Group III | –Originated at optic foramen<br>–Arachnoid plane (+/−) | | | |

Note: (−) negative involvement, (+) positive involvement, (+/−) may or may not be involved

## Answers

1. *What is your diagnosis and how would you grade the lesion?*
   - The lesion location and its MRI characteristics are mostly compatible with ACM which is a benign tumor originating from the cap-cells of the meninges covering the anterior clinoid process (ACP).
   - Cushing and Eisenhardt first coined the term in 1938[1] as part of medial sphenoid wing meningiomas.
   - Later, it was classified as a separate entity into three groups by Al-Mefty[2,3] (▶ **Table 10.1**).
2. *How do you explain the clinical findings in this case, based on the radiographic and anatomical observations?*
   - Tumor extension medially or inferiorly may lead to visual impairment and varying degrees of visual field defects due to involvement of the optic nerve or optic chiasm.
   - Superior growth into the posterior frontal lobe can give rise to seizures or Foster–Kennedy syndrome with anosmia, optic nerve atrophy on the same side, and contralateral papilledema.
   - Inferior extension into the cavernous sinus or along the tentorial edge may produce double vision due to cranial nerve III palsy, facial hypoesthesia, or ophthalmoplegia.
   - Finally, posterior and lower extension may encase the internal carotid artery, middle cerebral artery and its branches, and could lead to ischemia.
3. *How do you determine the site of origin for larger meningiomas in these regions?*
   - The definite site of larger meningiomas in the vicinity of this region is difficult to identify preoperatively. However, the anterior clinoid as a site would be supported by the presence of hypertrophied ACP on imaging, the epicenter of the tumor residing more on the ACP, the direction of tumor growth being predominantly superior laterally or posteriorly oriented toward the sylvian cistern, and the pattern of growth with greater than two-thirds of the tumor component lying outside the cavernous sinus.
   - On the other hand, a tumor engulfing the tuberculum as its epicenter with bone hyperostosis and displacement of the optic chiasm posteriorly and optic nerves laterally will support the diagnosis of tuberculum sellae meningioma.
   - Diaphragma sellae meningiomas may grow supra- or intrasellar with displacement of the pituitary stalk or compression of the optic chiasma as in pituitary tumor.
4. *What are your management goals, treatment options, and future follow-up plans?*
   - The goals of treatment are to decompress the optic nerve and achieve gross total resection of the tumor. The extent of tumor resection plays a key factor in minimizing tumor recurrence in the patient's lifetime.
   - A larger tumor with encasement of major intracranial vessels or involvement of the cavernous sinus represents a great surgical challenge and may defy the ambition of gross total resection. A subtotal safe removal of the tumor complemented with stereotactic radiotherapy may be a satisfactory treatment option in certain cases. Periodical follow-up with imaging is recommended.
5. *What are the important investigations and medical management measures prior to surgery, that are specific in the case of this lesion?*

   Preoperative preparations include the following:
   - Ophthalmology and endocrine consultation
   - CT scan of the head to evaluate hyperostosis
   - MR angiography (MRA), CT angiography (CTA), or digital subtraction angiography (DSA) to evaluate vascular involvement or to perform embolization in selected cases
   - Treatment with steroids and anticonvulsant should be initiated.
   - Intraoperative monitoring including cranial nerves, brainstem evoked potentials, and somatosensory evoked potentials (SSEP) should be ordered.
   - Surgical adjuncts, such as neuronavigation, microscope and ultrasonic aspirator, should be made available.
   - Microsurgical instruments such as Rhoton Micro dissectors, high-speed drill, rongeurs and curettes, and self-retaining retractors should be made available.
   - A lumbar drain can be considered in case of smaller- or moderate-sized tumors to reduce brain turgor intraoperatively.
   - The abdomen and/or thigh should be prepared for possible fat graft harvesting.

## Answers (*continued*)

6. *What are the generally recommended steps in the neurosurgical treatment of these lesions?*
   Pearls for safe microsurgical resection of these tumors include:
   - Performing a tailored skull base approach with resection of the sphenoid wing, flattening of the orbital roof. One may also consider orbital rim osteotomy or use of a FOZ approach for the removal of a larger tumor in order to enhance exposure, minimize brain retraction, and provide multiple surgical avenues to the lesion (▶**Fig. 10.2**).
   - Application of the “five Ds” concept (▶**Fig. 10.3**):
     - Deroof/decompress the optic nerve: deroof the optic nerve to facilitate its early identification, decompression, and safe manipulation, and to retrieve tumor extension in the foramina.
     - Drill the sphenoid wing, and the ACP (+/-): drill the ACP via an intradural or extradural approach, if needed, for better exposure (using a diamond burr with profuse irrigation). Neuronavigation is a valuable tool for anatomical localization especially during this step.
     - Devascularize the tumor: devascularize the tumor by interrupting its basal supply from the recurrent meningeal artery, the inferolateral trunk, or the tentorial artery.
     - Decompress the tumor: decompress the tumor centrally or segmentally starting from its base.
     - Dissect the tumor capsule: dissect the tumor capsule from the brain and maintain the integrity of the arachnoid membrane using irrigation and dynamic retraction if needed. One should respect the perforators during dissection and preserve them. Doppler ultrasound is a valuable tool to localize the encased vessels.
7. *How would you handle bleeding from the cavernous sinus intraoperatively?*
   - Bleeding from the cavernous sinus can be handled by using fibrin glue (Tisseel) or oxidized cellulose (Surgicel) and applying gentle pressure with a cottonoid and the suction catheter.
8. *What are the general outcomes after removal of this tumor?*
   - Optic canal involvement can be seen in more than one-third of clinoidal meningiomas with rates of improvement or stabilization of visual acuity being in the order of 90%.
   - Tumor recurrence rates are dependent upon extent of resection and tumor type, and are in the order of 10%.
   - Tumor progression rates after subtotal resection can be up to 30 to 40%.
9. *What are some postsurgical considerations when faced with an inability to totally remove the tumor?*
   - We recommend unroofing the optic canal to assure decompression of the optic nerve in all cases even if radiological infiltration is not demonstrated.
   - When total removal of the tumor is not possible, decompressing the optic pathways optimizes the administration of radiosurgery to the residual tumor, with lower risk to the optic apparatus.

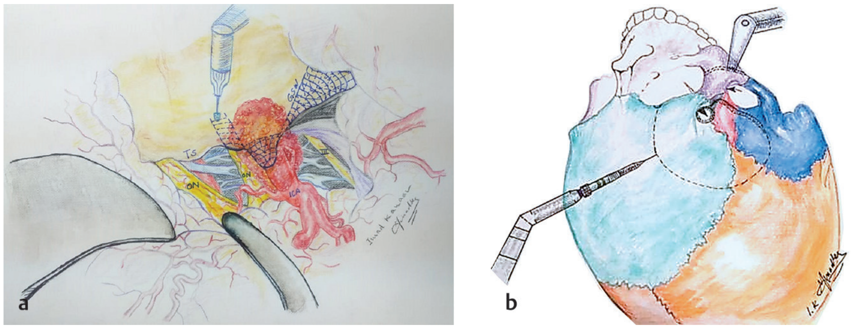

**Fig. 10.2** (**a**) Artistic illustration of the fronto-orbito-zygomatic approach (FOZ). Arrow head: McArthurs's burr-hole, arrow: to inferior orbital fissure, dashed line: frontotemporal craniotomy including the orbital rim part of the zygoma. (**b**) Artistic illustration of right anterior clinoid meningioma surrounding the internal carotid artery and compressing the right optic nerve. the dashed line represents bone removal of the sphenoid wing, anterior clinoid process, & the roof of the optic nerve canal.

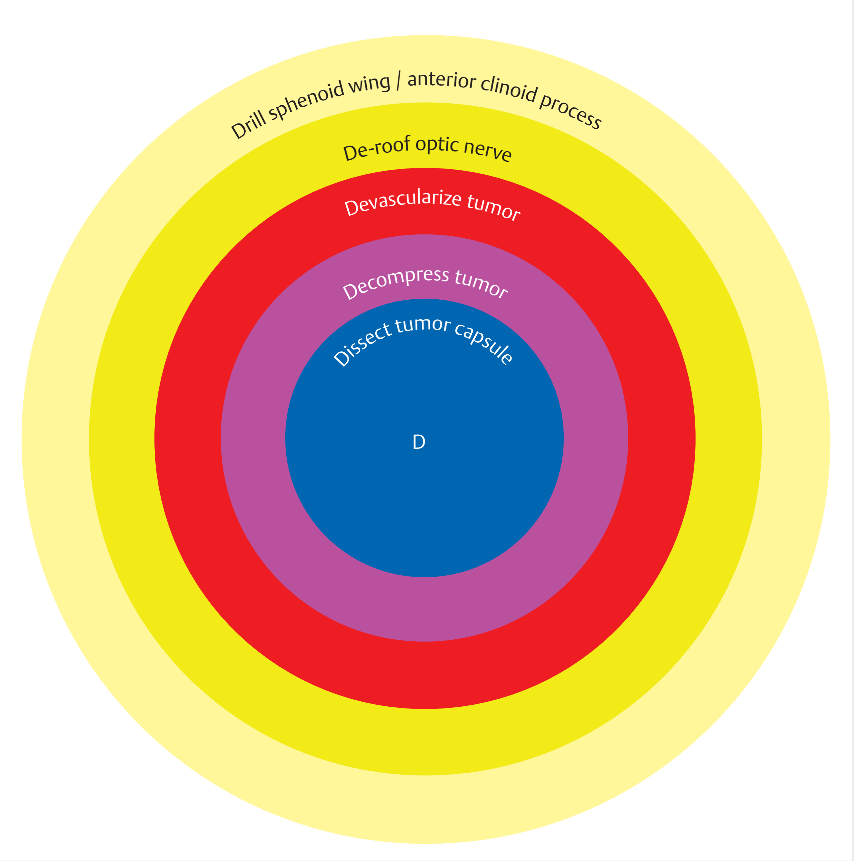
Drill sphenoid wing / anterior clinoid process
De-roof optic nerve
Devascularize tumor
Decompress tumor
Dissect tumor capsule
D

**Fig. 10.3** The "5 Ds" concept in removing a clinoidal meningioma.

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# Case 11 Velum Interpositum Meningioma

*Michel W. Bojanowski and Denis Klironomos*

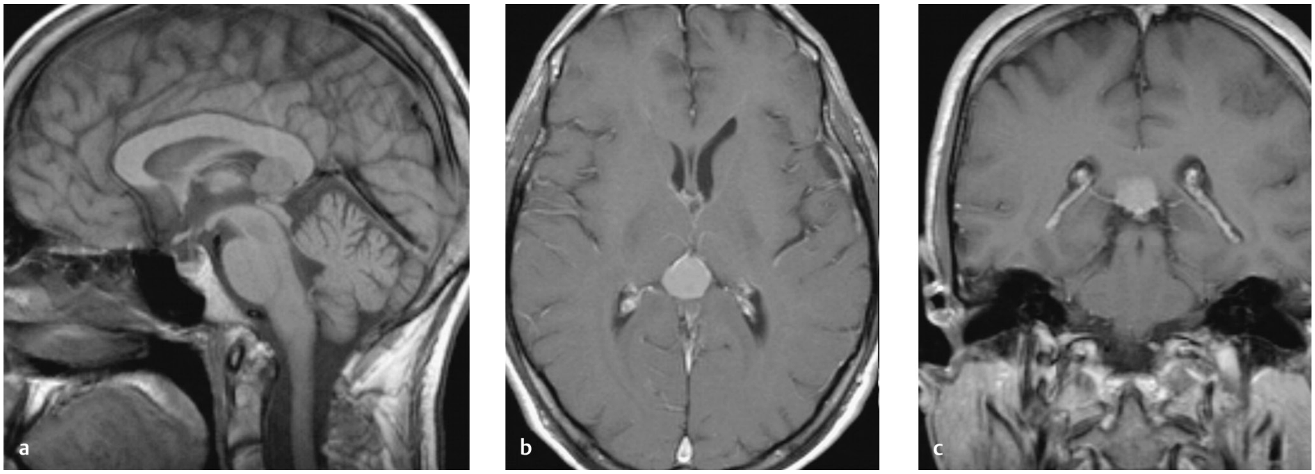

**Fig. 11.1** (**a**) Sagittal, (**b**) axial, and (**c**) coronal T1-weighted magnetic resonance images with gadolinium.

## Clinical Presentation

- A 40-year-old man previously in good health presented with a history of a nonprogressive headache ongoing for 1 year. There was no nausea, vomiting, or visual disturbance.
- MRI scan is shown in ▸**Fig. 11.1**.

## Questions

1. Interpret the MRI.
2. What is the differential diagnosis?
3. What additional studies would you like to order?

   You decide to obtain tissue for histo-pathological analysis.
4. What are the possible surgical approaches?
5. Describe the regional anatomy surrounding the pineal gland.
6. What are the potential surgical complications?

   The postoperative MRI (▸**Fig. 11.2**) reveals complete resection of the tumor. The histo-pathological study is diagnostic for a World Health Organization grade I transitional meningioma.
7. Explain the presumed origin of this tumor.
8. What are the reported locations of meningiomas without dural attachment?

## Answers

1. *Interpret the MRI.*
   - There is a well-circumscribed, round mass in the pineal region, isointense to the cortex with homogeneous enhancement after gadolinium injection. This lesion does not seem to originate from the pineal gland, which is compressed by the tumor.
   - There is no mass effect on the tectal plate or on the aqueduct of Sylvius and no hydrocephalus. The internal cerebral veins are pushed downward and consequently are beneath the tumor. The mass does not have any relation or attachment to the falcotentorial junction.
2. *What is the differential diagnosis?*
   - Meningioma: In the pineal region, meningiomas usually arise from tentorium cerebelli and falx. Few cases described without dural attachment are located in the velum interpositum.[1–3]
   - Pineoblastoma: Most are large, greater than 3 cm with peripheral calcifications. Usually associated with obstructive hydrocephalus.

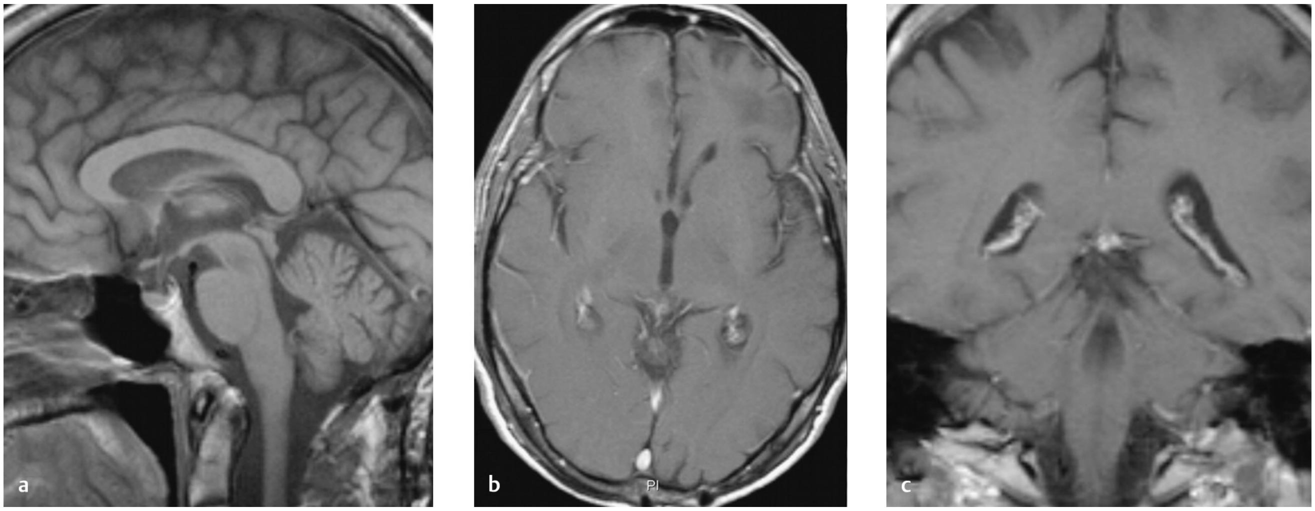

**Fig. 11.2** Postoperative (**a**) sagittal, (**b**) axial, and (**c**) coronal T1-weighted magnetic resonance images with gadolinium.

## Answers (*Continued*)

  - Pineocytoma: An enhancing, well-circumscribed pineal tumor with calcifications that rarely extends into the third ventricle.
  - Germ cell tumors: Engulf the pineal gland. Hyperintense to grey matter
  - Astrocytoma: Arises from midbrain tectum or thalamus; rarely from the pineal gland
  - Ependymoma: Mild to moderate heterogenous enhancement
  - Metastases

3. *What additional studies would you like to order?*
   - As neuroimaging alone is not consistently diagnostic for third ventricular meningioma, these lesions are usually evaluated according to standard algorithms for pineal masses.
   - Alfa-fetoprotein and beta human chorionic gonadotropin are markers of germ cell malignancy and should be measured in serum and cerebrospinal fluid if possible because patients with elevated markers suggestive of germinomas can be treated with chemotherapy and radiation without histological diagnosis.
   - For patients with previous history of malignancy, a complete metastatic workup should be done.
   - Consider a spinal survey MRI.
4. *What are the possible surgical approaches?*
   - Stereotactic-guided biopsy
     - Ideally suited for patients with contraindications to open surgery and general anesthesia. Also used for tumors that clearly invade the brainstem.[3]
     - However, it provides limited amount of tissue from lesions that may be histologically diverse.
     - The potential of hemorrhage is increased in the pineal region compared with other locations.[4]
     - Open surgical resection
     - The selection of surgical approach is determined according to:
       - The relationship of the tumor to the deep venous system and other surrounding structures
       - Particular characteristics of the tumor (size, spread, etc.)
       - Degree of surgeon's familiarity
   - Approaches include (▶**Fig. 11.3**)
     - Infratentorial supracerebellar approach
       - For tumors that displace the internal cerebral veins dorsally
       - Tumor is reached through the midline, below the deep cerebral veins
       - Avoids violation of normal tissues
     - Occipital transtentorial approach
       - For lesions above the deep venous system, midline, or above the tentorial edge
       - With this approach, it is difficult to dissect the tumor from the tela choroidea of the third ventricle
       - Necessitates retraction of the occipital lobe
     - Posterior transcallosal approach
       - For lesions anterior to the confluence of the deep cerebral veins
       - For lesions that displace the internal cerebral veins ventrally
   - Because of its relatively small size, the tumor was approached through an infratentorial supracerebellar corridor, avoiding violation of normal tissues.
5. *Describe the regional anatomy surrounding the pineal gland.*
   - The pineal gland is attached to the posterior wall of the third ventricle and projects posteriorly in the quadrigeminal cistern.
   - The splenium of the corpus callosum lies above this region and the thalamus is located on each side.

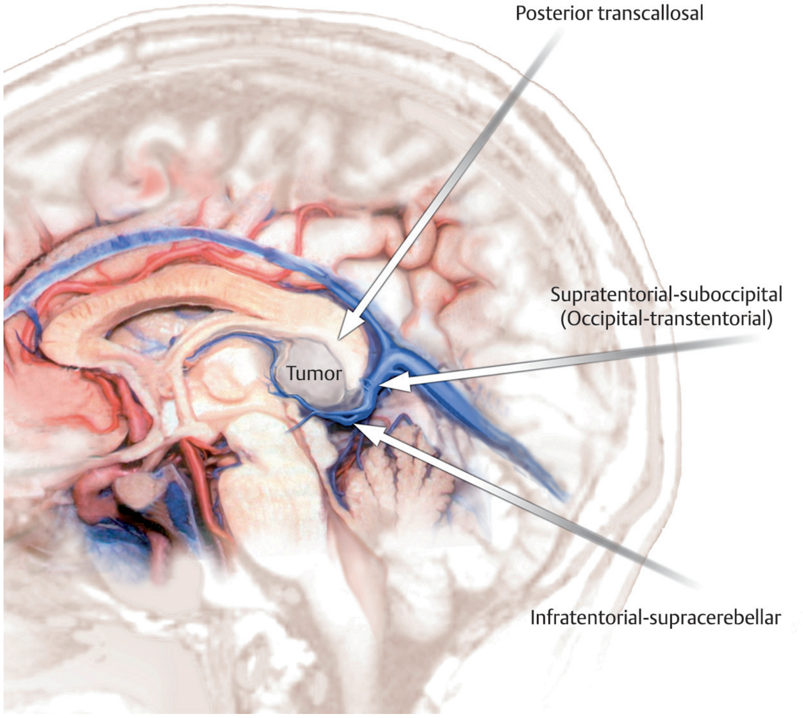
Posterior transcallosal
Supratentorial-suboccipital (Occipital-transtentorial)
Tumor
Infratentorial-supracerebellar

**Fig. 11.3** Surgical approaches to the pineal region.

## Answers (*continued*)

- The roof of the third ventricle is formed by the body and crura of the fornices and by the dorsal and ventral layers of the tela choroidea. The space between these two layers is the velum interpositum. The internal cerebral veins and the medial posterior choroidal arteries course through this space.
- The vein of Galen is located behind the posterior wall of the third ventricle.[5]

6. *What are the potential surgical complications?*
   - Oculomotor deficits including Parinaud's syndrome
   - Injury of the deep venous system, for example, venous occlusion leading to venous infarction or venous sinus tear leading to profuse bleeding
   - Venous air embolism related to the sitting position
   - Hemisensory or motor deficit (brain retraction), venous cortical infarction, and disconnection syndrome when using the transcallosal approach
   - Visual-fields deficit; venous cortical infarction when using the transtentorial approach
7. *Explain the presumed origin of this tumor.*
   - This case is a meningioma of the third ventricle without dural attachment.
   - Meningothelial cells are normally found in the arachnoid and choroidal tela. During embryological development, arachnoid tissue migrates together with the choroid plexus as the ventricular system invaginates, and thus meningocytes are found in the stroma of the choroid plexus.[6]
   - Ventricular meningiomas may arise from choroid plexus, the tela choroidea, the tania fornices, or the connective tissue of the pineal body.[3,6]
   - The velum interpositum is the potential space between the dorsal and ventral layers of the tela choroidea.[5,7] In the present case, it is presumed that the meningioma originated from arachnoid cap cells of the dorsal tela choroidea of the third ventricle with ventral displacement of the internal cerebral veins.[2]
8. *What are the reported locations of meningiomas without dural attachment?*
   - Meningiomas rarely occur in cerebral ventricles.
   - They are more commonly seen in the atrium of the lateral ventricle and for unknown reasons more frequently on the left side.[6] They have also been found in the third ventricle and only a few cases have been reported in the fourth ventricle.[6,8]

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# Case 12 Pituitary Apoplexy

*Michel W. Bojanowski and Denis Klironomos*

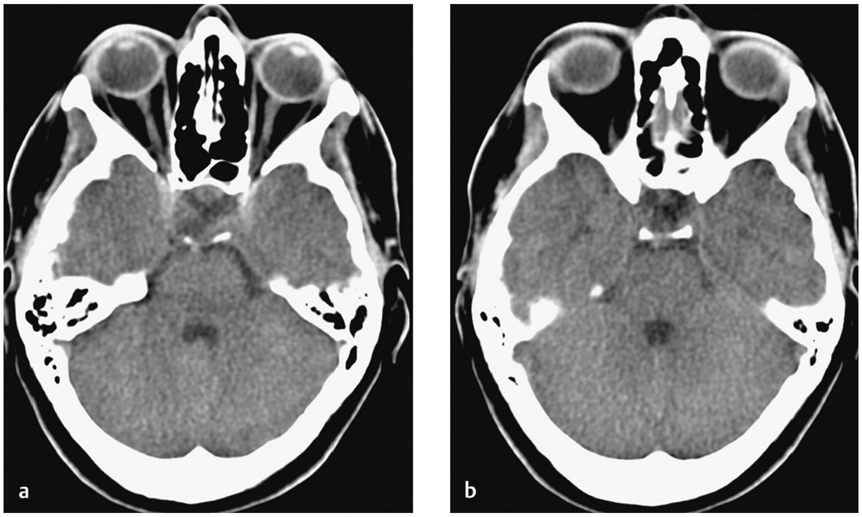

**Fig. 12.1** (**a, b**) Head CT scan showing axial cuts through the sella turcica.

## Clinical Presentation

- A 46-year-old woman with no previous medical history comes to you after awakening from a sudden headache associated with nausea and vomiting. She also had temporary visual loss which subsided spontaneously after 1 hour. She currently has a persistent severe headache.
- Upon examination, she is slightly obese. Blood pressure is 110/80 mm Hg. The neurological and neuro-ophthalmic examinations are within normal limits.
- ▶**Fig. 12.1** shows the CT scan of the head.

## Questions

1. Interpret the CT scan.
2. What is your initial diagnosis and why?
3. What are the important points to look for during the patient's history and physical examination?
4. What is your initial management?
   The headache has gradually decreased over the next few hours. An MRI scan of the brain is obtained (▶**Fig. 12.2**).
5. Describe the MRI (▶**Fig. 12.2**). What is your management now?
6. What are the indications for surgery?
   A follow-up MRI is obtained a few weeks later (▶**Fig. 12.3**).
7. Discuss the findings (▶**Fig. 12.3**).
8. What is the pathophysiology of this condition?
9. What are the precipitating factors?
10. What is the expected outcome after appropriate management?

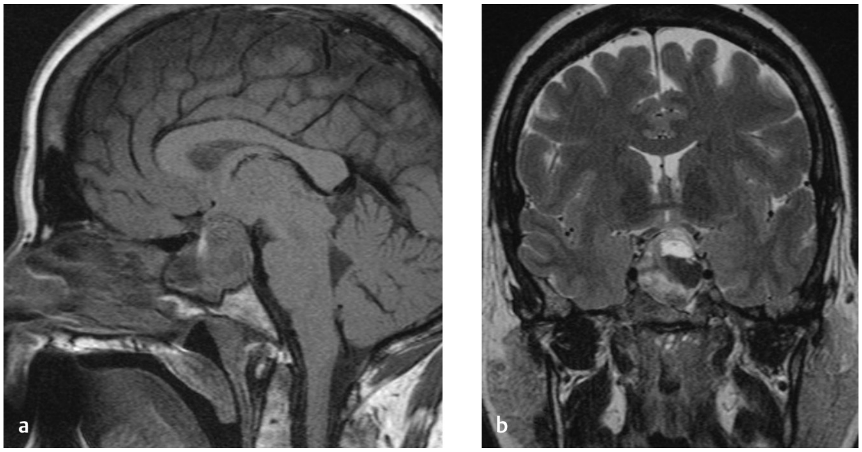

**Fig. 12.2** (**a**) Sagittal T1-weighted and (**b**) coronal T2-weighted weighted MRI of the brain at the level of the sella.

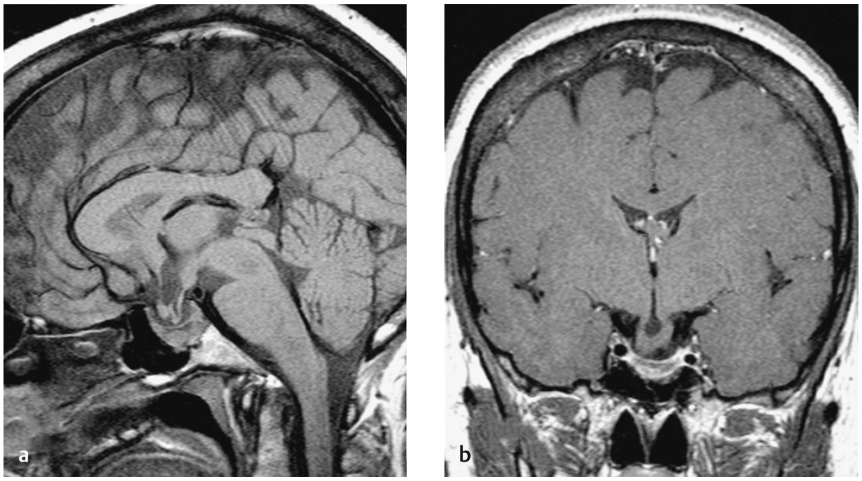

**Fig. 12.3** (**a**) Sagittal T1-weighted MRI scan without gadolinium and (**b**) coronal T1-weighted MRI scan with gadolinium at the level of the sella.

## Answers

1. *Interpret the CT scan.*
   The CT scan reveals significant enlargement of the pituitary fossa suggestive of an intrasellar tumor. There is no evidence of subarachnoid blood.
2. *What is your initial diagnosis and why?*
   - Pituitary apoplexy (PA)
   - This condition typically presents with sudden onset evolving within hours to 1 or 2 days. It includes a severe headache associated with nausea, vomiting, and/or a decreased level of consciousness. It may be accompanied by impairment of visual acuity, restriction of visual fields, ophthalmoplegia, and hormonal deficits.[1–3] It is caused by hemorrhage or necrosis of the pituitary gland. For some authors, PA refers to a clinical syndrome, thus, asymptomatic hemorrhage or necrosis of the pituitary gland is not considered as PA.[1–5]
   - Often PA is the first presentation of a pituitary tumor.[5]
3. *What are the important points to look for during the patient's history and physical examination?*
   - Acute severe headache with/without nausea accompanied or not with vomiting
   - Symptoms and signs suggestive of hypocorticism resulting from destruction or compression of the pituitary gland
   - Impairment of visual acuity and visual fields, resulting from superior extension of the tumor, and ophthalmoplegia when the sudden enlargement of the tumor is lateral toward the cavernous sinus.
   - Meningismus due to leakage of blood and necrotic tissue in the subarachnoid space
   - Precipitating factors (*refer to Question 9*).
   - Symptoms and signs related to the presence of a secreting or nonsecreting (hypopituitarism) pituitary tumor
   - PA may mimic more common pathological conditions. Differential diagnosis most commonly involves subarachnoid hemorrhage, followed by meningitis.[1,4-6]

## Answers (*continued*)

4. *What is your initial management?*
   - Hypopituitarism is present in the majority of patients who present with apoplexy[1,2] and requires the maintenance of hemodynamic stability, electrolyte balance, and rapid initiation of steroid replacement (intravenous hydrocortisone), after having blood samples drawn for baseline endocrine function tests to avoid an adrenal crisis.[5]
   - Immediate lab work, including cortisol, prolactin, luteinizing hormone, follicle-stimulating hormone, thyroid-stimulating hormone, T3, T4, and insulin-like growth factor-1
   - Urgent MRI to confirm the suspected diagnosis, view the underlying pathology, and rule out the possibility of a ruptured aneurysm
   - Ophthalmologic consultation
   - Endocrinology consultation to assist in management
   - Urgent surgical decompression is not necessary if the patient is neurologically intact with no visual impairment[4,5]
5. *Describe the MRI (* ▶**Fig. 12.2***). What is your management now?*
   - The MRI (▶**Fig. 12.2**) reveals a pituitary tumor with a suprasellar extension containing mixed intensities, suggestive of an acute intratumoral hemorrhage.
   - Neurological and neuro-ophthalmologic examinations are normal, and a large part of the tumor appears necrotic. Hence, conservative management is justified: obtain follow-up MRI in a few weeks provided the patient remains stable.
6. *What are the indications for surgery?*
   - Opinions may vary among different authors.
   - Deteriorating level of consciousness.
   - Urgent decompression is required for sudden onset of blindness or for progressive deterioration of visual acuity or visual fields.[3–5]
   - For mild visual impairment, surgery is recommended within the first week.[3–5]
   - Ventricular drainage may be necessary in the presence of hydrocephalus.
7. *Discuss the findings.*
   The MRI (▶**Fig. 12.3**) reveals the complete disappearance of the tumor. Sporadic cases of PA cured by isolated medical treatment have been reported.[7]
8. *What is the pathophysiology of this condition?*
   - It results from hemorrhage, infarction, or hemorrhagic infarction of a pituitary tumor secondary to its rapid growth.[1] This may be due to a discrepancy between the rate of neoplastic progression and the availability of circulatory input. Also, high intrasellar pressure may play a role. However, although PA typically occurs in pituitary macroadenomas, small tumors also hemorrhage.[8] One theory suggests intrinsic vasculopathy of the pituitary adenoma with secondary susceptibility to infarction and hemorrhage.[7]
   - Most patients had an undiagnosed pituitary adenoma at the time of apoplexy presentation.[5,8] Although PA occurs most of the time in pituitary adenomas (most often nonfunctioning adenomas), it may also occur, though rarely, in:[1,2]
     - Healthy pituitary gland
     - Pituitary abscess
     - Metastatic tumor
     - Lymphocytic hypophysitis
     - Craniopharyngioma
9. *What are the precipitating factors?*
   Precipitating factors are identified in ~50% of cases.[9] Precipitating factors that have been involved include the following:[4,5,10]
   - Hypertension
   - Treatment with bromocriptine
   - Major surgery (e.g., cardiac, orthopedic)
   - Anticoagulation therapy
   - Pituitary stimulation tests
   - Coagulopathies
   - Head trauma
   - Pregnancy
10. *What is the expected outcome after appropriate management?*
    - PA is a potentially life-threatening condition, but the overall outcome with appropriate management is good.[6,11]
      - Visual acuity: outcome is related to duration, severity of the initial defect, appearance of the optic disc, and timing of decompression.[1,11] However, even complete blindness may have remarkable improvement if surgical decompression is undertaken early.[12]
      - Ophthalmoparesis is reported to have good outcome whether treated conservatively or with surgical decompression.[7]
      - Endocrine function: the majority of patients require endocrine replacement therapy.[1,3,7,11]
    - Thickening of the sphenoid sinus mucosa may be an indirect measure of increased intrasellar pressure. This finding has been associated with higher grades of apoplexy, larger tumors with compression of parasellar structures, and worse endocrinological and neurological outcomes.[13]
    - Recurrent apoplexy has been documented in patients managed conservatively after their first apoplectic event, but it is rare after surgical treatment of the initial episode.[3]
    - Patients need to be followed closely for recurrence of tumor growth.[4]

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# Case 13 Secreting Pituitary Lesion

*Mohammed Alghamdi, Diana Ghinda, and Fahad AlKherayf*

## Clinical Presentation

- A 51-year-old woman presents with a 3-year history of weight gain, generalized muscle weakness, and facial hair growth.
- On examination, her blood pressure is 160/80 mm Hg and body mass index (BMI) is 32. The patient has central fat distribution and facial hair.

## Questions

1. What is the differential diagnosis and what are the features of Cushing's syndrome?
2. Given the results below, what is your diagnosis? And how do you confirm the diagnosis of Cushing's syndrome?
   - Cortisol am 682 nmol/L
   - Cortisol pm 383 nmol/L
   - 24-hour urine collection 699 nmol/L
   - After 1 mg dexamethasone (DST) suppression test, cortisol at 8 am is 450 nmol/L
3. What are the causes of Cushing's syndrome and what is the most likely cause in this patient given the following investigation?
   - High adrenocorticotropic hormone (ACTH)
   - High-dose DST suppression test leads to suppression of the morning serum and urine cortisol
4. An MRI with and without contrast was performed. Please describe the findings (▶**Fig. 13.1**).
5. Results of inferior petrosal sinus sampling (IPSS) 5 minutes post corticotropin-releasing hormone (CRH) stimulation are below. What is your interpretation?
   - Peripheral ACTH 15.1 pmol/L
   - Right IPSS ACTH 66.4 pmol/L
   - Left IPSS ACTH 15.7 p mol/L
6. Outline the management at this stage?
7. What are the possible complications occurring in the postoperative period?
8. The patient's symptoms did not improve and cortisone and ACTH are still elevated. IPSS still localizes to the right. What are the next options?
9. Multiple transsphenoidal surgeries were performed but failed. What do you recommend for this patient?
10. The patient underwent total bilateral adrenalectomy (TBA). Four months after the procedure she presented with hyperpigmentation.
    a. What is the most likely diagnosis?
    b. Discuss the pathophysiology of this condition.
    c. How would you manage this patient?

## Answers

1. *What is the differential diagnosis and what are the features of Cushing's syndrome?*
   Differential diagnosis includes: Cushing's syndrome, obesity, eating disorders, diabetes mellitus, alcoholism, polycystic ovarian syndrome, obstructive sleep apnea, major depression, and generalized anxiety disorder.[1]

   The clinical and endocrinological features encountered in the Cushing's syndrome are:[2,3]
   - General: obesity and hypertension
   - Skin: hirsutism, plethora, purple striae, acne, and bruising
   - Neuromuscular: lethargy, weakness (proximal myopathy), and osteopenia
   - Gonadal: menstrual irregularities, decreased libido, and impotence
   - Metabolic: diabetes mellitus and glucose intolerance, hyperlipidemia, polyuria, and kidney stones
   - Psychiatric: emotional liability, depression, euphoria, and psychosis
2. *Given the results above, what is your diagnosis? And how do you confirm the diagnosis of Cushing's syndrome?*
   The diagnosis is Cushing's syndrome. The first step to confirm the diagnosis is to exclude exogenous steroid use that can be identified by reviewing the patient's history and medications. If patient is not on steroids, perform one or more of the following tests:[3,4]

## Answers (*continued*)

- Low-dose DST suppression test: The patient is given 1 mg of DST at bedtime (11 pm); the following morning the plasma cortisone is measured (at 8 am). Normally the AM cortisone levels will decrease because of the negative feedback of DST on ACTH production; however, in patients with Cushing's syndrome, the feedback is lost. The test is positive if the plasma level of cortisone is more than 1.8 µg/dL (50 nmol/L). If less than 1.8 µg/dL, the test is negative for Cushing's syndrome.
- 24-hour urine collection for urine-free cortisone: normally urine cortisone is less than 135 nmol/24 hours; however, patients with Cushing's syndrome will have a high urine cortisone.
- Late night salivary cortisone (minimum of two measurements): normally cortisone levels are highest in the morning and decrease over the day, this is known as diurnal rhythm. In Cushing's syndrome, the diurnal rhythm is lost, so cortisone levels are high in the morning and late at night. The salivary samples are taken between 11 pm and midnight. The test is positive if salivary cortisone is more than 4 nmol/L.

3. *What are the causes of Cushing's syndrome and what is the most likely cause in this patient given the above investigation results?*
   The causes of Cushing's syndrome can be ACTH dependent or independent.[3]
   - ACTH-dependent etiologies include: Cushing's disease (most common cause) and ectopic ACTH-secreting tumors
   - ACTH-independent etiologies: iatrogenic, adrenal neoplasm, and nodular adrenal hyperplasia
   - Since ACTH is elevated and the high-dose DST suppressed cortisone levels, the most likely diagnosis is Cushing's syndrome.
4. *An MRI with and without contrast was performed. Please describe the findings (▶* **Fig. 13.1***).*
   Coronal T1- and T2-weighted MRI show a small nodule within the pituitary gland more to the right side that could represent a pituitary microadenoma. T1 sagittal and axial cuts with gadolinium did not show clear enhancement of the nodule.
5. *Results of inferior petrosal sinus sampling (IPSS) 5 minutes post corticotropin-releasing hormone (CRH)*

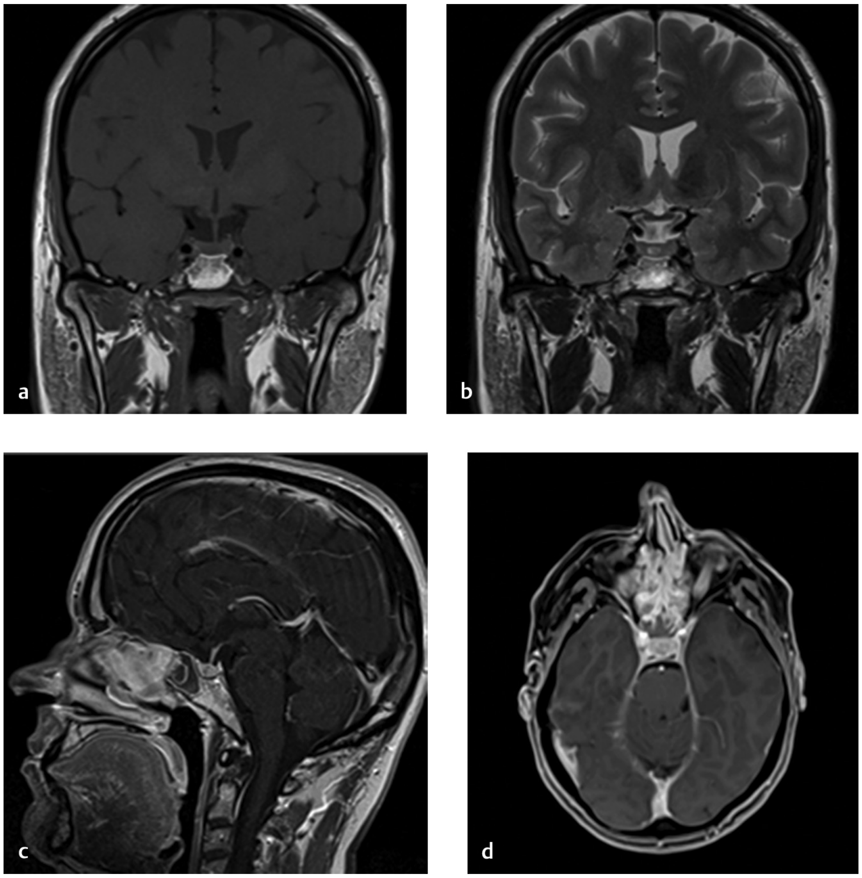

**Fig. 13.1** MRI coronal views (**a, b**) and axial (**c**) and sagittal (**d**) contrasted views.

## Answers (*continued*)

*stimulation are listed above. What is your interpretation?*
- ACTH levels are two times higher in the IPSS compared to the peripheral blood sample, which is consistent with Cushing's disease.
- ACTH levels are elevated in the right IPSS but not in the left IPSS (the ratio is more than 2), this localizes the lesion to the right side.
- Explanation: The pituitary gland drains into the cavernous sinus which drains into the inferior petrosal sinus and subsequently into the jugular vein. ACTH levels in both petrosal sinuses and peripheral blood are measured before and 5 minutes after CRH stimulation. If ACTH levels are two times higher in the IPSS compared to the peripheral blood, the test is considered positive for pituitary adenoma. IPSS has a high diagnostic accuracy reaching 100% in expert hands.[3] Also, IPSS can help localizing the side of the tumor if one side shows higher levels of ACTH compared to the contralateral side.

6. *Outline the management at this stage?*
   Endonasal transsphenoidal resection of the ACTH-secreting adenoma is considered the treatment of choice. The remission rates achieved vary from 65 to 90% for microadenomas and from 50 to 65% for macroadenomas.[5] Medical management can be used as a preoperative temporizing measure and an option for nonsurgical candidates or patients in whom surgery and/or radiotherapy have failed.[6] Preoperative optimization of hormonal excess or insufficiency is an important step prior to surgery. A detailed review of the patient's imaging should be performed with a particular attention to the sellar floor, parasellar sinuses, and surrounding structures including the internal carotid arteries, pituitary gland and stalk, and the optic apparatus.
7. *What are the possible complications occurring in the postoperative period?*
   Neuroendocrine insufficiency, hematoma, epistaxis, ischemic events, hydrocephalus, cerebrospinal fluid leaks, visual changes, or meningitis. In terms of endocrine issues, it is common to encounter diabetes insipidus (DI) in the early postoperative period as well as hyponatremia and excessive urine output (250 cc/hour for 2–3 hours).[7] The cortisol function is also reassessed in the postoperative period and if there is evidence of hypocortisolemia (cortisol < 10 mcg/dL), glucocorticoid replacement is indicated.[8]
8. *The patient's symptoms did not improve and cortisone and ACTH are still elevated. IPSS still localizes to the right. What are the next options?*
   - Recurrence rates after surgery are high and can occur also in a delayed fashion. Medical management, radiation, or repeat surgery represent the potential management options in these cases.
   - A repeat pituitary surgery is an option that can result in the cure of Cushing's disease in approximately 50% of cases.[9] Radiotherapy can also represent an alternative option but can take months to years to attain the full effect and can cause panhypopituitarism.[10]
   - Medical management includes drugs that act by inhibiting steroidogenesis in the adrenal glands with ketoconazole and metyrapone being the most common drugs employed. Other potential medications are multireceptor ligand somatostatin analogs, second-generation dopamine agonists, or glucocorticoid receptor antagonist.
9. *Multiple transsphenoidal surgeries were performed but failed. What do you recommend for this patient?*
   - Bilateral adrenalectomy represents the next surgical option which usually results in resolution of hypercortisolemia. Nonetheless, this option causes glucocorticoid and mineralocorticoid deficiency with Nelson's syndrome occurring in up to 35% of these patients.[11,12] Although bilateral adrenalectomy represents an effective and rapid-onset alternative, corticotroph adenoma progression is seen in about 30% of cases.[11]
10. *The patient underwent total bilateral adrenalectomy (TBA). Four months after the procedure she presented with hyperpigmentation.*
    a. *What is the most likely diagnosis?*
    b. *Discuss the pathophysiology of this condition.*
    c. *How would you manage this patient?*
    a. Nelson's syndrome is the most likely diagnosis.
    b. The pathophysiology relates to the fact that the negative feedback from cortisone is lost which can lead to enlargement of the preexisting pituitary adenoma leading to increased ACTH secretion which stimulates melanocytes to synthesize melanin leading to hyperpigmentation.
       The following features confirm the diagnosis of Nelson's syndrome in a patient who underwent bilateral adrenalectomy for the treatment of refractory Cushing's disease.[13]
       – Radiological evidence of an enlarging pituitary lesion
       – High ACTH levels (more than 500 ng/L)
       – An increase ACTH of more than 30% compared to postoperative ACTH levels
    c. Surgical resection is the first-line management option, especially in the presence of mass effect on the optic apparatus. If surgery failed to control the disease, radiotherapy could be used. Medical management with selective somatostatin analogue, sodium valproate, dopamine agonist, and temozolomide has also been described.[13]

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# Case 14 Nonfunctioning Pituitary Adenoma

*Michael S. Taccone, Hubert Lee, John Woulfe, and Fahad AlKherayf*

## Clinical Presentation

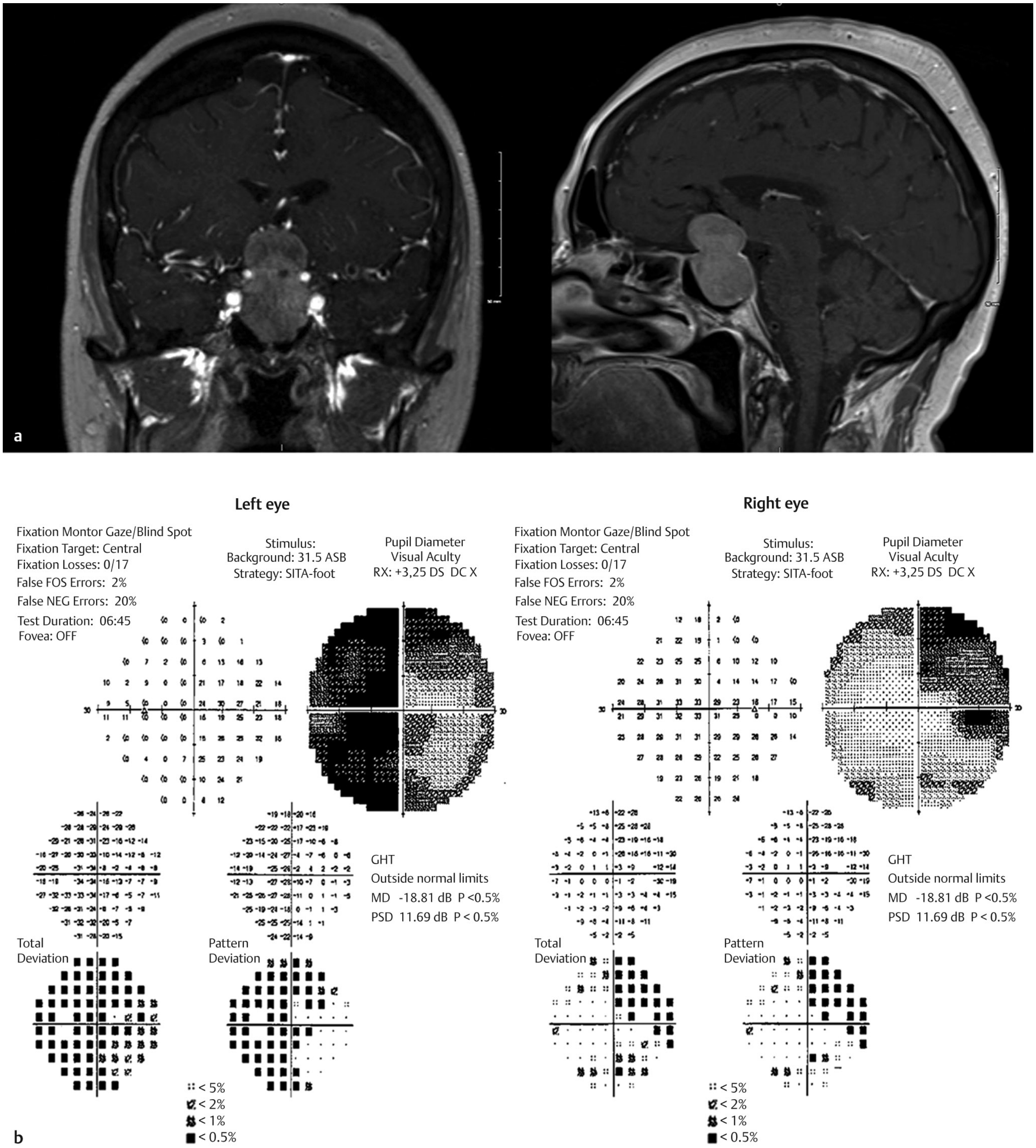
- A 55-year-old previously healthy female presents with progressive difficulty appreciating her blind spot while driving.
- She presented to her family physician who found bilateral temporal visual field loss on confrontation during an otherwise normal physical exam.
- Subsequently, she was sent to your neurosurgical referral service for further management.

## Questions

1. What is the differential diagnosis for sellar masses?
2. What are the necessary investigations for a patient with a suspected pituitary mass?
3. ▶**Fig. 14.1** and ▶**Table 14.1** show the investigations you ordered, interpret the results.
4. What is your diagnosis?
5. What is the role of surgery in nonfunctional adenomas?
6. Describe your surgical approach/exposure to this lesion.
7. List the intracranial and intranasal complications of endonasal resection of this tumor.
8. You are drilling laterally within the sella and note a brisk flow of blood. Determine the possible complication and describe your management.
9. Your patient returns for a postoperative visit complaining of a persistent runny nose. Determine the possible complication and describe your management.
10. Identify the structures in ▶**Fig. 14.2a** and describe the histological slide in ▶**Fig. 14.2b**.

## Answers

1. *What is your differential diagnosis for sellar masses?*
   - Broad differential for sellar masses includes:
     - Anterior pituitary tumors
     - Rathke's cleft cysts
     - Meningioma
     - Arachnoid cyst
     - Vascular lesions
     - Metastases
     - Inflammatory and vasculitides
     - Infectious causes[1]
   - A landmark study by Famini et al identified the most common etiologies of sellar masses in 2,598 patients who underwent pituitary MRI. Of patients with adenomas, 40% had prolactinomas, 37% had nonfunctional adenomas, and 13% had growth-hormone-secreting adenomas. Whereas 18% of patients had nonadenomatous masses of which 19% were Rathke's cleft cysts, 15% were craniopharyngiomas, and another 15% were meningiomas.
2. *What are the necessary investigations for a patient with a suspected pituitary mass?*
   - This patient presents with signs and symptoms suspicious for a sellar mass with optic chiasmal compression. Thus, preliminary investigations must aim to achieve following three goals:
     - An anatomic diagnosis via imaging
     - Assessing the hormonal impact or functionality of the lesion, if any, via biochemical analysis
     - Quantifying the loss of visual function and acuity due to mass effect on the optic nerve via formal visual field testing
   - The ideal imaging modality which allows for the greatest resolution and detail is a dedicated T1-weighted coronal view thin slice (2–3 mm thick) pituitary MRI with gadolinium contrast agent.[2] MRI imaging is helpful to delineate the relationship of the tumor to the pituitary gland, parasellar, and suprasellar vascular and parenchymal structures for operative planning.
   - If the relationship between the adenoma and vascular structures of the cavernous sinus or circle of Willis is needed, a 3D time-of-flight MR angiography (MRA) should be obtained.
   - Also, CT scan to investigate the relationship of the bony skull base to the sella turcica, internal carotid arteries, and cavernous sinus can be helpful.


a
Left eye
Fixation Montor Gaze/Blind Spot
Fixation Target: Central
Fixation Losses: 0/17
False FOS Errors: 2%
False NEG Errors: 20%
Test Duration: 06:45
Fovea: OFF
Stimulus:
Background: 31.5 ASB
Strategy: SITA-foot
Pupil Diameter
Visual Acuity
RX: +3,25 DS DC X
GHT
Outside normal limits
MD -18.81 dB P <0.5%
PSD 11.69 dB P < 0.5%
Total Deviation
Pattern Deviation
Right eye
Fixation Montor Gaze/Blind Spot
Fixation Target: Central
Fixation Losses: 0/17
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False NEG Errors: 20%
Test Duration: 06:45
Fovea: OFF
Stimulus:
Background: 31.5 ASB
Strategy: SITA-foot
Pupil Diameter
Visual Acuity
RX: +3,25 DS DC X
GHT
Outside normal limits
MD -18.81 dB P <0.5%
PSD 11.69 dB P < 0.5%
Total Deviation
Pattern Deviation
< 5%
< 2%
< 1%
< 0.5%
b

**Fig. 14.1** (**a**) MRI with contrast of the brain. (**b**) Visual field evaluation.

**Table 14.1** Laboratory values including endocrine panel of patient described herein

| Parameter | Level | Normal Values |
|---|---|---|
| Prolactin | 49 ng/mL | Nonpregnant females (2–29 ng/mL) |
| Cortisol | 123 nmol/L | 83–607 nmol/L |
| Luteinizing hormone | 22 IU/L | Menopause (14–52 IU/L) |
| Follicle-stimulating hormone | 40 IU/L | Menopause (25–134 IU/L) |
| Growth hormone | 3 ng/mL | 1–8 ng/mL |
| Thyroid-stimulating hormone | 1.37mIU/L | 0.30–5.60 mIU/L |
| Free T4 | 6.5 mcg/dL | 4.5–11mcg/dL |
| Free T3 | 120 ng/dL | 100–200 ng/dL |
| Serum sodium | 142 mmol/L | 135–145 mmol/L |
| Serum osmolarity | 292 mmol/kg | 280–300 mmol/kg |
| Serum potassium | 4.6 mmol/L | 3.5–5.0 mmol/L |

## Answers (*continued*)

- Although, pituitary macroadenomas are typically hormonally silent, mass effect on the infundibulum may result in tertiary hypopituitarism. Thus, a full screening hormone panel (prolactin, growth hormone, luteinizing hormone, follicle-stimulating hormone, cortisol, thyroid-stimulating hormone, a-subunit, thyroxine, cortisol, insulin-like growth factor type 1, testosterone, and estradiol) is required.
- Furthermore, if hypercortisolism (Cushing's disease) is suspected, sampling of the inferior petrosal sinus may be necessary.
- Finally, urgent referral to a neuro-ophthalmologist must be made for formal visual acuity and visual field testing. This serves as not only a tool to direct urgency of surgical intervention and decompression but also as a baseline assessment of visual function to assist with monitoring of recovery and potential complications postoperatively such as an expanding hematoma.

3. ▶**Fig. 14.1** *and* ▶**Table 14.1** *show the investigations you ordered, interpret the results.*
   - The MRI in ▶**Fig. 14.1a** demonstrates a large 2 × 3 cm mass within the sella turcica. There appears to be a large superior suprasellar extension of the mass with compression of the optic apparatus.
   - An anatomic explanation for the patient's symptoms has been found; however, two critical steps in the work-up of this patient follow. An endocrinological work-up and a formal assessment of the visual fields preoperatively.
   - ▶**Table 14.1** displays a normal hormone panel with normal serum electrolytes and osmolarity with the exception of an elevated prolactin. However, this does not suggest a functional prolactinoma, rather the elevated prolactin is due to impaired functioning of the hypothalamic–pituitary axis and reduced dopamine transmission to the anterior pituitary which inhibits prolactin secretion (a phenomenon known as "stalk effect").
   - Finally, the visual field report in ▶**Fig. 14.1b** demonstrates a bitemporal hemianopsia with a junctional scotoma.
4. *What is your diagnosis?*
   It is a nonfunctional pituitary macroadenoma.
5. *What is the role of surgery in nonfunctional adenomas?*
   - Since nonfunctional pituitary adenomas do not produce an excess of hormones, the typical presenting symptoms are not that of hypersecretory syndromes but rather the result of mass effect on the pituitary gland or neighboring structures.
   - These tumors are often macroadenomas with varying amounts of suprasellar and parasellar extension.
   - Surgical resection provides a mean to decompress these structures with greater success in improving headaches and symptoms attributable to optic chiasm compression than pituitary function.[3] The main goals of surgery are to relieve mass effect and cytoreduction to achieve adequate tumor control rather than complete tumor resection.
6. *Describe your surgical approach/exposure to this lesion.*
   - The patient is positioned supine on the operating table with the head rigidly fixated in neutral position, slightly rotated in the direction of the surgeon. A zero (0) degree endoscope is inserted into the right nostril to visualize the inferior and middle turbinates as well as the nasal septum. Neuronavigation may be used to assist in landmark identification. The choana can be found following the floor of the nasal cavity to the posterior aspect of the inferior turbinate. Lateralizing or removal of the middle turbinate will allow exposure of the sphenoid ostium typically 1 cm above the choana.
   - At this point, a nasoseptal flap may be elevated. An anterior sphenoidotomy is then preformed removing a portion of the vomer and sphenoid rostrum using a high-speed drill or Kerrison Bone Punches. Inferolateral extension should be limited to avoid injury to the sphenopalatine artery. The sphenoid mucosa and septum should then be visible and

## Answers (*continued*)

removed with care as the septum may overlie the internal carotid artery. At the end of this exposure, the endoscopic view seen in ▶**Fig. 14.2a** should be obtained.

- The sellar floor can then be opened using a high-speed drill to thin the bone and Kerrison Bone Punches exposing the dura mater. This can be incised using a scalpel with a telescopic blade to reveal the lesion.
- Location confirmation may be aided by use of neuronavigation or fluoroscopy at this point.

7. *List the intracranial and intranasal complications of endonasal resection of this tumor.*
   - Intracranial complications[4]
     - Cerebrospinal fluid leak
     - Meningitis
     - Anterior pituitary insufficiency
     - Intrasellar hematoma
     - Tension pneumocephalus
     - Cranial nerve injury
     - Diabetes insipidus
     - Internal carotid artery injury/pseudoaneurysm
   - Intranasal complications[5]
     - Prolonged nasal crusting
     - Synechiae formation
     - Septal deviation
     - Epistaxis
     - Sinusitis
8. *You are drilling laterally within the sella and note a brisk flow of blood. Determine the possible complication and describe your management.*
   - The source of bleeding is likely the internal carotid artery which lies lateral to the sella within the cavernous sinus. Prevention through knowledge of anatomy (i.e., preoperative imaging) and meticulous dissection are the best means of hemostasis and avoidance.
   - In the event of an arterial injury, categorizing into low flow (small perforating vessels) or high flow (a major artery such as the internal carotid artery) can help determine the appropriate technique to employ.[5]
   - For high-flow arterial bleeds, suction should be used to direct bleeding away from the endoscope lens to aid visualization to identify the site of vascular injury. Pressure with a pledget and a second suction can be used to achieve a dry field. Bipolar electrocautery is then applied to the vessel side wall to seal the defect. If a seal is difficult to obtain, one should use hemostatic agents such as fibrin glue combined with collagen along with cotton pledgets. Intraoperative or early postoperative angiography should be considered to rule out and treat potential vessel occlusion or pseudoaneurysm.
   - In the case of carotid injury, the anesthetist should be informed immediately for maintenance of the patient's blood pressure and intravascular volume. Neurophysiologic monitoring, if available, can act as a correlate for cerebral perfusion. If hemostasis cannot be achieved intraoperatively, the site should be packed and the patient taken emergently to the angiography suite for endovascular stenting or vessel occlusion.
   - Low-flow arterial bleeds can be managed with bipolar cauterization or focal packing with hemostatic agents (i.e., microfibrillar collagen) in areas where vessel retraction is not a possibility.
9. *Your patient returns for a postoperative visit complaining of a persistent runny nose. Determine the possible complication and describe your management.*
   - Persistent postoperative nasal discharge should raise the suspicion of cerebrospinal fluid (CSF) leakage. This can be tested by having the patient lean forward for several minutes. The emitted fluid should be sent for beta-2 transferrin testing, a protein found exclusively in CSF.
   - Postoperative CSF leaks can be managed with bed rest and continuous lumbar drainage for 3 or more days or surgically through endoscopic endonasal exploration and repair.
   - Lumbar drainage, while conservative, has the risk of overdrainage, pneumocephalus, meningitis, and lengthening hospital stay.[6] One can monitor for pneumocephalus and meningitis via neuroimaging and serial clinical neurological examination within the first 24 to 48 hours of therapy.
   - Failure to achieve resolution of CSF leakage after 72 hours warrants consideration of surgical repair particularly in the setting of high-flow CSF leakage and pneumocephalus.[7]
   - When the site of CSF leakage is difficult to localize, high-resolution CT and CT cisternography can be used to identify bony defects and demonstrate active leaks, respectively.[5] Intraoperative localization can be assisted with intrathecal fluorescein although reported complications include seizures, postprocedural headache, nausea, vomiting, dizziness, tinnitus, opisthotonus, cranial neuropathy, and lumbar radiculopathy.[8]
10. *Identify the structures in* ▶**Fig. 14.2***a and describe the histological slide in* ▶**Fig. 14.2***b*.

    In ▶**Fig. 14.2a**, an intraoperative endoscopic view of the sphenoid sinus is shown. The lateral optocarotid recess (A), the optic nerve (B), the paraclival segment of the internal carotid artery (C), the sellar floor (D), and the planum sphenoidale (E) are shown.

    ▶**Fig. 14.2b** shows a hematoxylin and eosin stain slide of a nonfuntional pituitary macroadenoma. There is a higher than expected degree of cellularity without an abundance of mitotic figures. There is also loss of the normal reticulin network characteristic of normal anterior pituitary architecture.

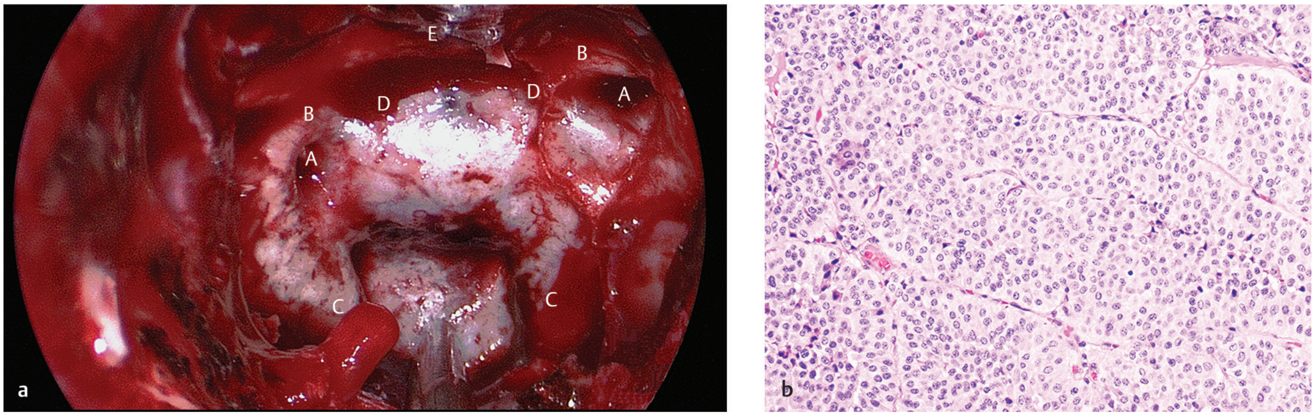

**Fig. 14.2** (**a**) Intraoperative endoscopic picture and (**b**) pathological section.

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# Case 15 Craniopharyngioma: Endoscopic Approach

*Daniel M. Prevedello, Amin B. Kassam, André Beer-Furlan, and Ricardo L. Carrau*

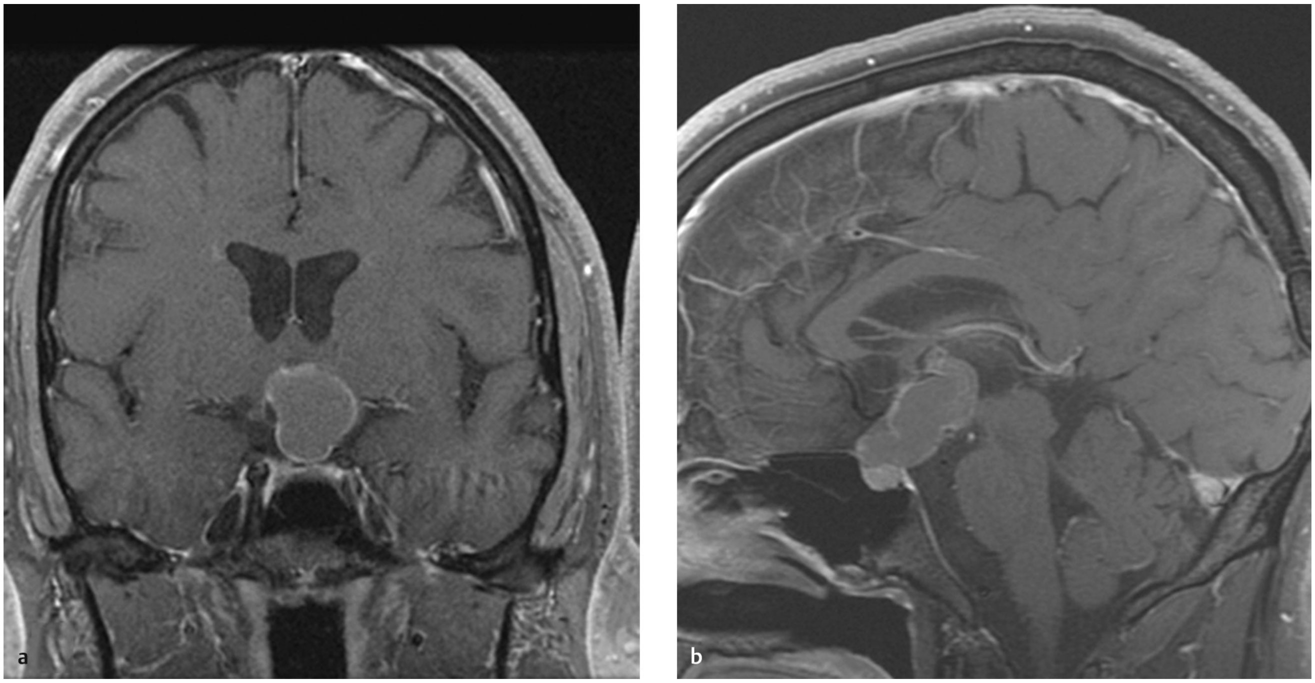

**Fig. 15.1** (**a**) Coronal and (**b**) sagittal T1-weighted magnetic resonance images of the brain with contrast showing a suprasellar mass.

## Clinical Presentation

- A 44-year-old woman presents with mild decreased peripheral vision, increased thirst, and fatigue.
- A recent diagnosis of hypothyroidism was given by her primary care physician.
- MRI scan is shown in ▶**Fig. 15.1**.
- The patient also underwent a CT scan of the brain which does demonstrate calcifications at the level of the lesion (*not shown here*).

## Questions

1. What is the diagnosis?
2. What is the differential diagnosis?
3. What are the treatment options?
4. What are the classic surgical approaches for craniopharyngiomas and their main limitations?
5. How are craniopharyngiomas classified in relation to the infundibulum? Which type is demonstrated in this case?

You decide to approach the tumor via an endoscopic endonasal route.

6. Describe the advantages of an endoscopic endonasal approach (EEA) over the standard transcranial approaches.
7. What are the limitations for EEAs for the treatment of craniopharyngiomas?
8. What are the prognostic factors for risks of recurrence or regrowth?

## Answers

1. *What is the diagnosis?*
   - Craniopharyngioma is most likely diagnosis: the diagnosis is made on the basis of both radiologic and clinical findings.
   - The contrast-enhanced MRI demonstrates a heterogeneous enhancing sellar and suprasellar lesion with solid and cystic components.
   - In addition, CT images confirm the presence of calcifications in the lesion, which are present in ~ 80% of craniopharyngioma cases.
   - Clinically, the presentation of panhypopituitarism and occasionally diabetes insipidus combined with these imaging studies is diagnostic for craniopharyngioma.
2. *What is the differential diagnosis?*
   - The differential diagnosis for suprasellar enhancing lesions includes (see Case 14 on Pituitary Adenoma)[1] craniopharyngioma, Rathke's cleft cyst, gliomas, germinoma, pituitary adenoma, meningioma, metastases, brain abscess, aneurysm, sarcoid, and teratoma.
3. *What are the treatment options?*
   - The main treatment modality for craniopharyngiomas is surgical resection. A total surgical resection decreases the possibility of a tumor recurrence. The first surgical attempt is the most important in determining the outcome because reoperation on a recurrent or residual craniopharyngioma is less likely to result in a complete resection due to scar tissue formation caused by the previous surgery or radiation.[2]
   - Other modalities of treatment, in general reserved for recurrent tumors, include:
     - Radiation (radiosurgery, intensity modulated radiation therapy, or fractionated radiotherapy)[2,3]
     - Stereotactic cystic drainage (with or without Ommaya reservoir placement)
     - Stereotactic intracystic delivery of radioactive or immunoactive substances such as:
       - Bleomycin[4]
       - Radioactive substances
       - Phosphorous-32 (P32)[5]
       - Yttrium-90 (Y90)[6]
       - Rhenium-186 (Re186)[7]
       - Interferon-α[8]
4. *What are the classic surgical approaches for craniopharyngiomas and their main limitations?*
   - Frontal interhemispheric and pterional transsylvian approaches[9]:
     - This may include orbitozygomatic osteotomies and other variations.
     - The cranial–caudal angle of approach has a substantial limitation with the optic apparatus positioned between the surgeon and the target.
     - To improve corridors around the parachiasmatic space, critical positioning of the chiasm needs to be considered via these approaches, specifically whether the chiasm is positioned anteriorly (prefixed) or posteriorly (postfixed).
     - Anterior interhemispheric approach with subsequent opening of the lamina terminalis:
       - This is an option for tumors eroding the floor of the third ventricle. It is a midline approach well indicated to resect a midline lesion; however, it is very limited in the exposure of the undersurface of the optic apparatus and is usually indicated only for intraventricular craniopharyngiomas.
   - Transcortical or transcallosal through a transchoroidal approach to the third ventricle
     - It is a great option for isolated large tumors confined to the third ventricle.
   - Lateral presigmoid combined with a transtentorial subtemporal approach:
     - Option to minimize optic apparatus manipulation is a caudal–cranial angle of attack
     - Allows for exposure of the prepontine and interpeduncular cisterns
     - Nevertheless, any dissection through this lateral view is divided into many small corridors in between the cranial nerves (II to XII) that are present in the midline cisterns' lateral walls.
5. *How are craniopharyngiomas classified in relation to the infundibulum? Which type is demonstrated in this case?*
   - The classification is summarized in ▶ **Fig. 15.2**.[10]
     - Type I is preinfundibular
     - Type II is transinfundibular (extending into the stalk) as in this case illustration
     - Type III is retroinfundibular (extends behind the gland and stalk)[11]
     - Type IV is isolated to the third ventricle and/or optic recess and may not be accessible via an endonasal approach.
     - Craniopharyngiomas that present on the third ventricle can be resected endonasally when they are an extension of the suprasellar component.[12]
6. *Describe the advantages of an EEA over the standard transcranial approaches.*
   - The main advantage of an endonasal corridor to approach craniopharyngiomas is the fact that it is a midline approach for a midline lesion. In contrast to the lateral approaches for the interpeduncular fossa, it offers a direct visualization of the midline without the need for cranial nerve dissection.[13]
   - It provides an adequate corridor to the infra- and supradiaphragmatic midline craniopharyngiomas, also giving the opportunity to manage lesions extending into the third ventricle chamber.[12]
   - At the same time, it offers a caudal–cranial angle of attack, allowing for better visualization of the inferior aspect of the optic apparatus and hypothalamus, which is essential for preservation of chiasmatic perforators during tumor dissection.[13]

## Answers (*continued*)

- When compared with standard transsphenoidal approach, which uses the same corridor with microscopic visualization, the endoscopic technique is proven to be superior by offering a wider visualization of the ventral skull base.[14,15,16]

7. *What are the limitations for EEA for the treatment of craniopharyngiomas?*
   - Lack of space and/or three-dimensional perception: Freedom of movements is improved by using two surgeons synergistically performing the surgery through both nostrils coupled with better depth perception generated by continuous movements of the camera.
   - Any extension of the lesion laterally beyond the optic nerves formally determines a limitation for an endoscopic endonasal resection of craniopharyngiomas. Lesions located in those territories have to be accessed through paramedian corridors (craniotomies) either as a complementary or an exclusive approach for resection of specific lesions.
   - Lack of surgeon's experience with the endoscope.[15,16]
8. *What are the prognostic factors for risks of recurrence or regrowth?*
   - Favorable prognostic factors for risk of recurrence/regrowth in craniopharyngioma include:[2,16]
     - Size (< 4 cm)
     - Favorable location
     - Complete surgical removal
     - Age (> 5 years carries favorable prognosis)
     - Absence of severe hypothalamic involvement

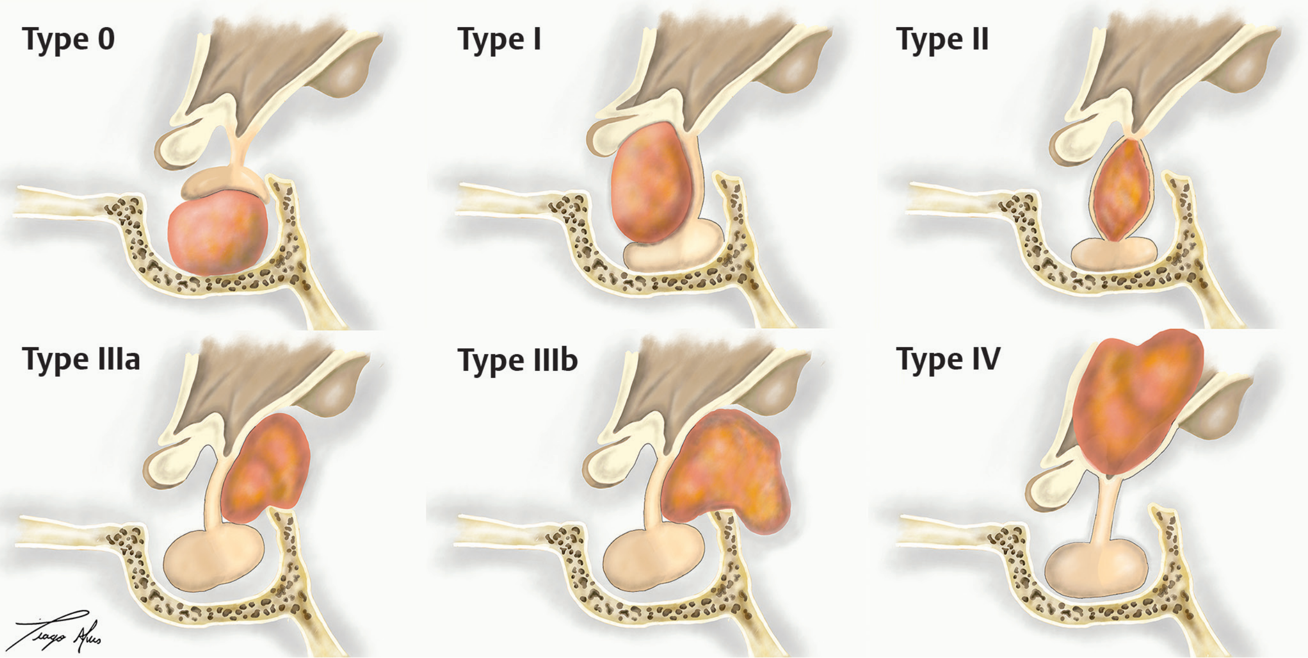

**Fig. 15.2** Artist's rendering of anatomic classification of craniopharyngiomas based on location with respect to the optic chiasm. See text for details (*Question 5*). Endoscopic endonasal approaches may not be ideal for resection of type IV craniopharyngiomas. However, these pure third ventricular lesions can be approached through a minimally invasive transcortical endoscopic approach. A port, which is a transparent cylinder with less than 12 mm of diameter, is inserted through the frontal cortex after dilatation of the cerebral tissue by an introducer. The dissection is performed under direct visualization of the ventricles and the lesion. (Source: Surgical Approach Selection. In: Stamm A, ed. Transnasal Endoscopic Skull Base and Brain Surgery: Surgical Anatomy and its Applications. 2nd Edition. Thieme; 2019.)

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# Case 16 Glioma—Surgical Treatment

*Mariam Alrashid, Khalid Bajunaid, and Kevin Petrecca*

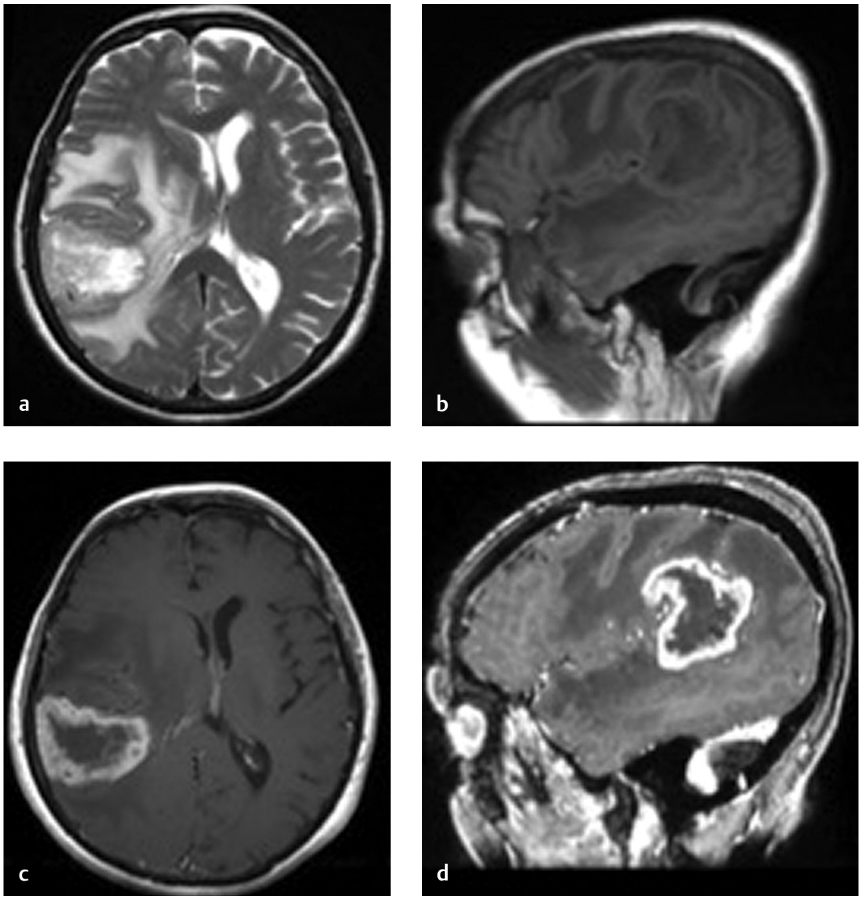

**Fig. 16.1** Brain MRI scan of patient described herein with axial T2-weighed image (**a**). Sagittal T1-weighted without contrast (**b**) and axial (**c**), and sagittal T1-weighted image with contrast (**d**).

## Clinical Presentation

- A 69-year-old woman presents to medical attention with difficulty in walking and left arm weakness. She was also complaining of a constant headache for the previous 3 days. Upon questioning, she reveals that while she could maintain her household, it was becoming increasingly difficult.
- On examination, she was found to be fully alert and oriented. Cranial nerve examination was normal. She had a partial left visual field defect, mild left hemiparesis, and gait imbalance.
- You have been consulted following the brain MRI (▶**Fig. 16.1**).

## Questions

1. Interpret the MRI brain.
2. Provide a relevant differential diagnosis.
3. What is the patient's Karnofsky Performance Status (KPS)?
4. What is your initial management?
5. What is the next step and what will you recommend for the patient?

You decide to operate and proceed with gross total resection. Her postoperative course is unremarkable. Two days after surgery, during your morning rounds, you notice that the patient is drowsy and less responsive than usual.

6. What are the possible causes of her deterioration?

## Questions (*continued*)

7. A CT scan of the head was unremarkable and the laboratory test revealed $Na^{2+}$ level of 126, how will you manage her now? What are the possible causes of the hyponatremia?
8. What adjuvant treatments would you recommend? Give the name of therapeutic agent(s) and doses.
9. The neuropathologist diagnosed the tumor as an isocitrate dehydrogenase (IDH) wild-type glioblastoma, what is the expected prognosis with your treatment?
10. ▶**Fig. 16.2** shows a potential residual tumor 3 months after treatment, how will you interpret this and how will you manage it?
11. Based on the Response Assessment in Neuro-Oncology (RANO), response criteria, how would you evaluate the treatment response?
12. What are the hypotheses proposed for the cell of origin of glioblastoma?

## Answers

1. *Interpret the MRI brain.*
   The MRI shows a ring-enhancing lesion in the right posterior temporal and parietal lobes. It is associated with extensive edema and a midline shift of 5 mm. The lesion is causing deformation of the ipsilateral ventricle.
2. *Provide a relevant differential diagnosis.*
   - Grade 4 IDH wild-type glioma (primary glioblastoma)
   - Secondary brain tumor (metastasis)
   - Abscess
   - Less likely diagnoses include:
     - Toxoplasmosis
     - Lymphomas
     - Resolving hematoma
     - Cysticercosis cyst
     - Traumatic brain injury
     - Infarction
3. *What is the patient's Karnofsky Performance Status (KPS)?*
   KPS is a performance status scoring system that estimates the functional capacity of cancer patients. The two most commonly used scoring systems are the KPS score and the Eastern Cooperative Group (ECOG) performance status score (▶**Table 16.1**).[1] The patient's KPS score is 70.
4. *What is your initial management?*
   - Admission to a well-monitored setting with hourly neurological assessment
   - Intravenous dexamethasone (10 mg) followed by 4 mg QID with antigastritis prophylaxis (either an H2 blocker or proton pump inhibitor)
   - No indication for antiepileptic prophylaxis if there is no history of seizure[2,3]
   - Complete blood cell count, electrolytes, coagulation profile, type and screen
   - Ophthalmology consultation to document the visual field defect
5. *What is the next step and what will you recommend for the patient?*
   - Discuss the following options with the patient:
     - Supportive care
     - Stereotactic biopsy to establish diagnosis
     - Surgical resection for diagnostic and therapeutic purposes
   - Given the patient's age, KPS, and tumor location, a safe gross total resection is feasible and recommended. Studies have shown that an extent of resection of more than 80% in glioblastoma multiforme (GBM) improves overall survival. Survival improves in a stepwise manner as the extent of the resection increases between 80 and 100%.[4–6]
6. *What are the possible causes of her deterioration?*
   - Intracranial hemorrhage at the surgical site
   - Meningitis
   - Diffuse worsening of edema secondary to venous congestion/infarction
   - Seizures
   - Hyponatremia
   - Hydrocephalus
   - Other systemic or cardiac event (pulmonary embolism, myocardial infarction, metabolic encephalopathy)
7. *A CT scan of the head was unremarkable and the laboratory test revealed $Na^{2+}$ level of 126, how will you manage her now? What are the possible causes of the hyponatremia?*
   - Monitor the urine output and order the following investigations to establish the cause of the hyponatremia: comprehensive metabolic panel, serum osmolality, urine osmolality and urine electrolytes.
   - Admit to intensive care unit for neurological observation
   - Correct the hyponatremia based on the latest guidelines:[7]
     - To avoid the risk of osmotic demyelination, the rate of increase in plasma sodium concentration should be less than 8 mmol/L over 24 hours with a maximum rise of 12 mmol/L. When treating patients at an increased risk of osmotic demyelination, as is the case for alcohol abusers, the target concentration should be less than 6 mmol/L with a maximum rise of less than 8 mmol/L.

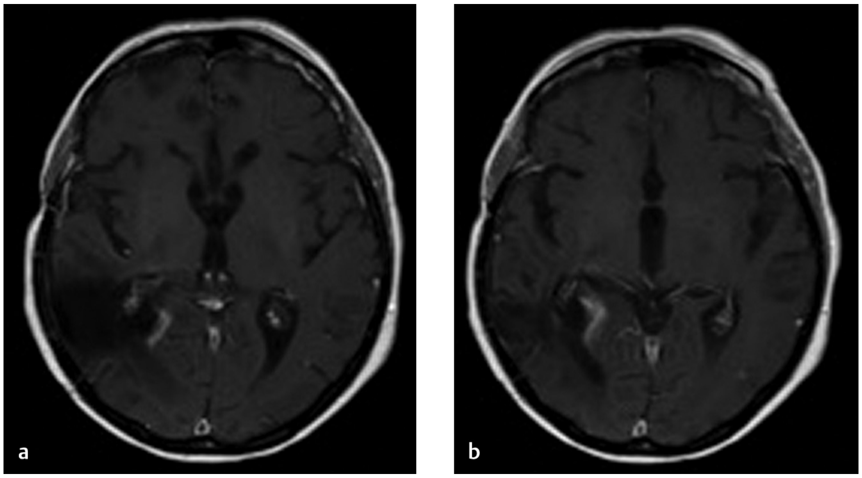

**Fig. 16.2** (**a, b**) Axial T1-weighted image with contrast performed 3 months after surgical resection.

**Table 16.1** Karnofsky grading scale of performance

| KPS | ECOG |
|---|---|
| 100: Normal activity, no evidence of disease | 0: Normal activity |
| 90: Normal activity with minor symptoms<br>80: Normal activity with effort (some symptoms) | 1: Ambulatory but restricted in physically strenuous activity |
| 70: Unable to perform normal activity but able to care for self<br>60: Requires occasional assistance | 2: Ambulatory > 50% of waking hours, capable of self-care, occasional assistance required |
| 50: Requires considerable assistance<br>40: Disabled, requires special assistance | 3: Ambulatory < 50% of waking hours, capable of only limited self-care, requires nursing support |
| 30: Severely disabled, hospitalization indicated<br>20: Hospitalization necessary, requires active supportive care<br>10: Moribund | 4: Bedridden |
| 0: Dead | 5: Dead |

*Abbreviations:* ECOG, Eastern Cooperative Group Performance Status; KPS, Karnofsky Performance Status.
*Source:* Data from Ma et al 2010.[1]

## Answers (*continued*)

- An initial increase of 3 to 5 mmol/L over 2 to 4 hours is recommended to reverse cerebral edema, reduce intracranial pressure, and prevent seizures. The remainder of the increase in plasma sodium can occur over 24 hours.

  The syndrome of inappropriate antidiuretic hormone secretion (SIADH) is the likely cause of the hyponatremia. Other causes of hyponatremia in the neurosurgical patient are: acute ACTH deficiency, cerebral salt wasting syndrome, and hypovolemia.

8. ***What adjuvant treatments would you recommend? Give the name of therapeutic agent(s) and doses.***
   The Stupp's protocol[8] includes is a combination of temozolomide and radiotherapy (RT) that is administered in the following manner:
   - RT: 2 Gy given 5 days per week for 6 weeks, for a total of 60 Gy
   - Concomitantly administer daily temozolomide 75 mg/m$^2$, 7 days a week from the first to the last day of RT
   - Six cycles of temozolomide (150–200 mg/m$^2$ once daily) on days 1 to 5 of a 28-day cycle beginning 4 weeks following completion of RT
9. ***The neuropathologist diagnosed the tumor as an IDH wild-type glioblastoma, what is the expected prognosis with your treatment?***
   - Progression-free survival is 6 months[9]
   - Median overall survival is 15 months in patients with an IDH wild-type GBM versus 31 months in patients with an IDH-mutant GBM[10]
   - Six-month progression-free survival rate: 41.0% in patients who undergo a gross total resection versus 21.1% in patients who undergo a subtotal resection[6]
10. ▶**Fig. 16.2** ***shows a potential residual tumor 3 months after treatment, how will you interpret this and how will you manage it?***
    - The most important differential at this stage, given the timeline, is pseudoprogression versus tumor progression.

## Answers (*continued*)

- Pseudoprogression[11,12] is a subacute postradiation reaction that manifests radiologically as a contrast enhancement, at the surgical cavity, on an MRI. It is a transient reaction that classically resolves spontaneously without treatment. It appears within 3 months after radiation treatment and mimics tumor recurrence on MRI.
- Pathophysiology: it is a transient interruption of myelin synthesis caused by radiation-induced injury to oligodendrocytes.
- Clinical presentation: most patients are asymptomatic but some might present with worsening of preexisting symptoms, transient cognitive decline, or somnolence syndrome.
- Treatment: close surveillance with serial imaging is usually sufficient, but for patients with symptoms and worsening edema corticosteroids are administered.
- If the serial imaging or other radiological modalities (▶**Table 16.2**) suggest a tumor recurrence or progression, then treatment is modified accordingly.
- A correlation has been observed between O6-methylguanine-DNA methyltransferase (MGMT) methylation status and pseudoprogression with 91% of MGMT-methylated tumors exhibiting pseudoprogression.

11. *Based on the Response Assessment in Neuro-Oncology (RANO), response criteria, how would you evaluate the treatment response?*
    - Based on the RANO criteria, the patient is considered to have a partial response.
    - The RANO response criteria, which is an updated version of the McDonald criteria, is an objective radiological and clinical assessment of treatment response and tumor recurrence for malignant gliomas (▶**Table 16.3**).[13]
12. *What are the hypotheses proposed for the cell of origin of glioblastoma?*

    There are two hypotheses:
    - Dedifferentiated glial cells that underwent genetic transformation
    - Malignant transformation of either a bipotential precursor cell or a neural stem cell (classically residing in the subventricular zone and the dentate gyrus of the hippocampus)[14]

**Table 16.2** Comparison of imaging characteristics: tumor recurrence versus radiation necrosis

| | MR Spectroscopy | Perfusion MRI | PET |
|---|---|---|---|
| Tumor | ↑ ↑ Cho/NAA<br>↑ ↑ Ch/Cr<br>↓ ↓ NAA/Cr<br>↓ NAA and Cr<br>↑ Cho and Lac | ↑ relative CBV (> 2.6 mL blood/g of tissue) | ↑ metabolic activity |
| radiation necrosis | ↑ Cho/NAA<br>↑ Ch/Cr<br>↑ NAA/Cr<br>↑ Cho | ↓ relative CBV (< 0.6 mL blood/g of tissue) | ↓ metabolic activity |

*Abbreviations:* Cho, choline (a cell membrane marker that is elevated in tumors and inflammatory processes); Cr, creatine plus phosphocreatine (a measure of energy stores); Lac, lactate (a product of anaerobic metabolism); NAA, N-acetyl aspartate (a neuronal marker that decreases with neuronal disease or loss of integrity).
Adapted from Hou LC, Veeravagu A, Hsu AR, Tse VC. Recurrent glioblastoma multiforme: a review of natural history and management options. Neurosurgical Focus 2006, 20(4):E5., and borrowed from Nader et al 2010.

**Table 16.3** Response assessment in neuro-oncology criteria

| **Response Type** | **The Criteria** (All the criteria must be fulfilled to establish a response except for disease progression) |
|---|---|
| Complete response | No new lesion in MRI images<br>No enhancing disease in T1W post-gadolinium MRI images (must be sustained for at least 4 weeks)<br>Stable or improved disease in T2W/FLAIR MRI images<br>Patient must be off corticosteroids (or on physiological replacement doses)<br>Patient is clinically stable or improved |
| Partial response | No new lesion in MRI images<br>≥ 50% reduction of enhancing disease in T1W post-gadolinium MRI images[a] (must be sustained for at least 4 weeks)<br>Stable or improved disease in T2W/FLAIR MRI images<br>Patient must be on the same or lower dose of corticosteroids<br>Patient is clinically stable or improved |
| Stable disease | No new lesion in MRI images<br>< 50% reduction and < 25% increase of enhancing disease in T1W post-gadolinium MRI images[a/b] (must be sustained for at least 4 weeks)<br>Stable or improved disease in T2W/FLAIR MRI images<br>Patient must be on the same or lower dose of corticosteroids<br>Patient is clinically stable or improved |
| Disease progression | New lesion in MRI images<br>≥ 25% increase in the enhancing disease in T1W post-gadolinium MRI images[b]<br>Significant increase in disease in T2W/FLAIR MRI images<br>Patient is on stable or increasing doses of corticosteroids<br>Patient is clinically deteriorating |

*Abbreviations:* FLAIR, fluid-attenuated inversion recovery; T1W, T1-weighted; T2W, T2-weighted.

[a] Compared with baseline in the sum of products of perpendicular diameters of all measurable enhancing lesions.

[b] Sum of the products of perpendicular diameters of enhancing lesions compared with the smallest tumor measurement obtained either at baseline (if no decrease) or best response.

*Source:* Data from Wen et al 2010.[13]

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# Case 17 Glioma—Epigenetics

*Mariam Alrashid, Khalid Bajunaid, and Kevin Petrecca*

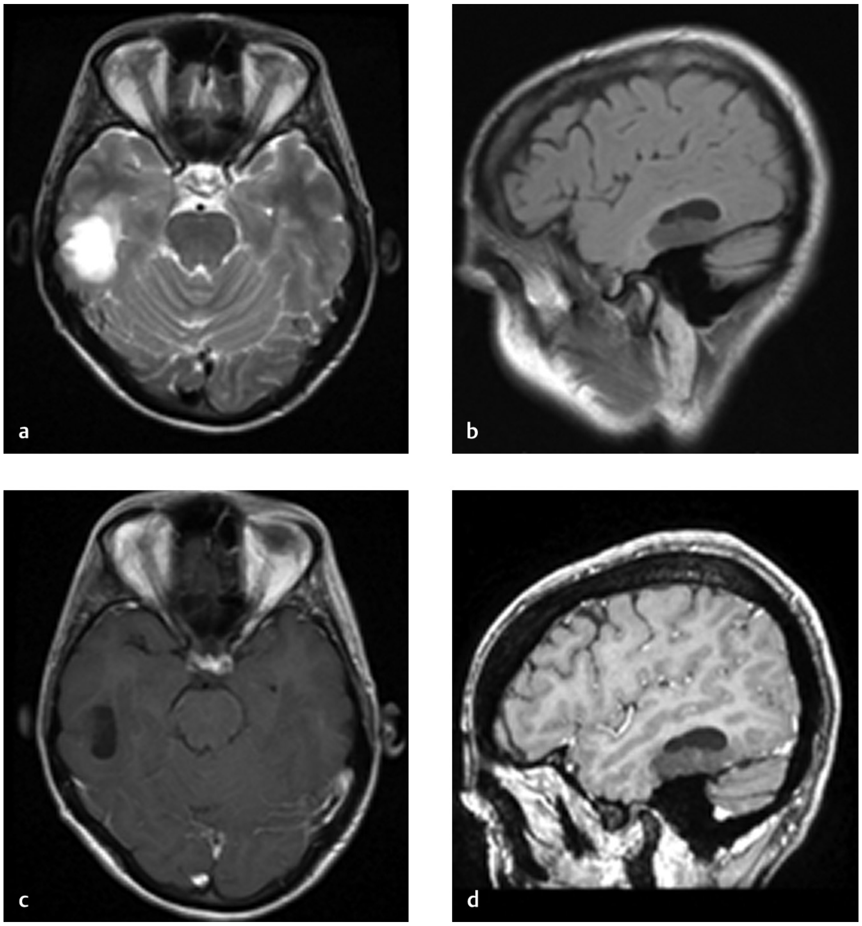


**Fig. 17.1** Brain MRI scan of patient described herein, with axial T2-weighed image (**a**), sagittal T1-weighted without contrast (**b**), and axial (**c**) and sagittal (**d**) T1-weighted image with contrast.

## Clinical Presentation

- A 49-year-old right-handed woman had a CT scan following a minor head trauma. The radiologist reported an incidental finding in the CT scan and recommended an elective brain MRI for further evaluation.
- The patient was referred to your clinic following the acquisition of the MRI (▶**Fig. 17.1**). When you interview her, she denies any symptoms and, on examination, she is neurologically intact.

## Questions

1. Interpret the MRI.
2. Provide a relevant differential diagnosis.
3. What is the natural history of each differential?
4. What is the 2016 WHO classification of gliomas?
5. What is your management plan?
   You advise her to proceed with surgical resection. The surgery and postoperative course were unremarkable. The final pathology report states the following:
   a. Diagnosis: oligodendroglioma grade 2
   b. Isocitrate dehydrogenase (IDH) 1: mutated and 1p19q co-deleted
   c. *ATRX* is present and the O6-methylguanine-DNA methyltransferase (MGMT) promotor is hypermethylated
6. How will the surgery impact her prognosis?
7. Following a complete resection of oligodendroglioma grade 2, what is the next step?
8. What is an IDH mutation? What is its significance in gliomas?
9. What does hypermethylation of the MGMT promoter mean? What is its significance? Four years following complete resection, the surveillance MRI is shown in ▶**Fig. 17.3**. Interpret the MRI.
10. What is the natural history of recurrent oligodendroglioma? What will you advise the patient?
    The cancer is completely resected. The pathologist diagnoses the tumor this time as a grade 3 oligodendroglioma. IDH 1 mutated and 1p19q co-deleted.
11. What is her prognosis in light of her tumor progression to a higher grade? What is your management plan?
12. What are the key genetic alterations in primary and secondary glioblastoma?

## Answers

1. *Interpret the MRI.*
   - There is a T2 hyperintense tumor in the right temporal lobe, mainly at the third temporal gyrus. This tumor does not exert significant mass effect.
   - The contrast-enhanced MRI does not show enhancement in or around the mass.
2. *Provide a relevant differential diagnosis.*
   - Astrocytoma (grade 2 or 3)
   - Oligodendroglioma (grade 2 or 3)
   - Ganglioglioma
   - Dysembryoplastic neuroepithelial tumor (DNT)
   - Pilocytic astrocytoma
3. *What is the natural history of each differential?*
   - Grade 2 and 3 gliomas: These tumors, despite their nomenclature as “low-grade gliomas,” are not benign tumors. Inevitably, they progress to a higher grade which may lead to death if not treated appropriately. The timing of malignant transformation of grade 2 gliomas is highly variable ranging between 2 to 10 years depending on the histopathological and molecular characteristics of the tumor.[1] The median overall survival of untreated grade 2 gliomas is just less than 6 years.[2]
   - Ganglioglioma: In most cases, this lesion has a very benign clinical course. However, malignant progression and secondary glioblastoma have been reported in some patients.[3–5] It is the most common tumor seen in young patients with chronic temporal lobe epilepsy and is associated with drug-resistant epilepsy.
   - DNT: This tumor has a benign course with hardly any potential for malignant transformation, but it is typically associated with chronic pharmaco-resistant epilepsy in up to 90–100% of patients.[6]
   - Pilocytic astrocytoma: This tumor has a benign course and it is usually limited in its infiltrative capacity, but there are rare documented instances of malignant transformation in adult patients.[7] If left untreated, it might eventually lead to neurological deficits or signs of intracranial hypertension based on its location.
4. *What is the 2016 WHO classification of gliomas?*
   The 2016 WHO classification stratifies gliomas primarily based on their IDH mutational genetic status while still incorporating the histological diagnosis into the classification (▶**Fig. 17.2**).
5. *What is your management plan?*
   - One should present the following options to the patient:
     - Stereotactic biopsy to establish diagnosis
     - Surgical resection for diagnostic and therapeutic purposes
     - “Watch and wait”: Close observation with regular MRIs of the brain to evaluate the evolution of the tumor with the endpoints being onset of symptoms or signs of radiological tumor progression. Although this approach was historically acceptable, it is no longer considered an appropriate option. Moreover, early surgical management of grade 2 gliomas has demonstrated a survival benefit as it minimizes the residual tumor burden. On the other hand, waiting for symptom onset or radiological changes runs the risk of a patient presenting with a more extensive disease and a lower Karnofsky

## Answers (*continued*)

Performance Score (KPS) that might impact their outcome negatively.

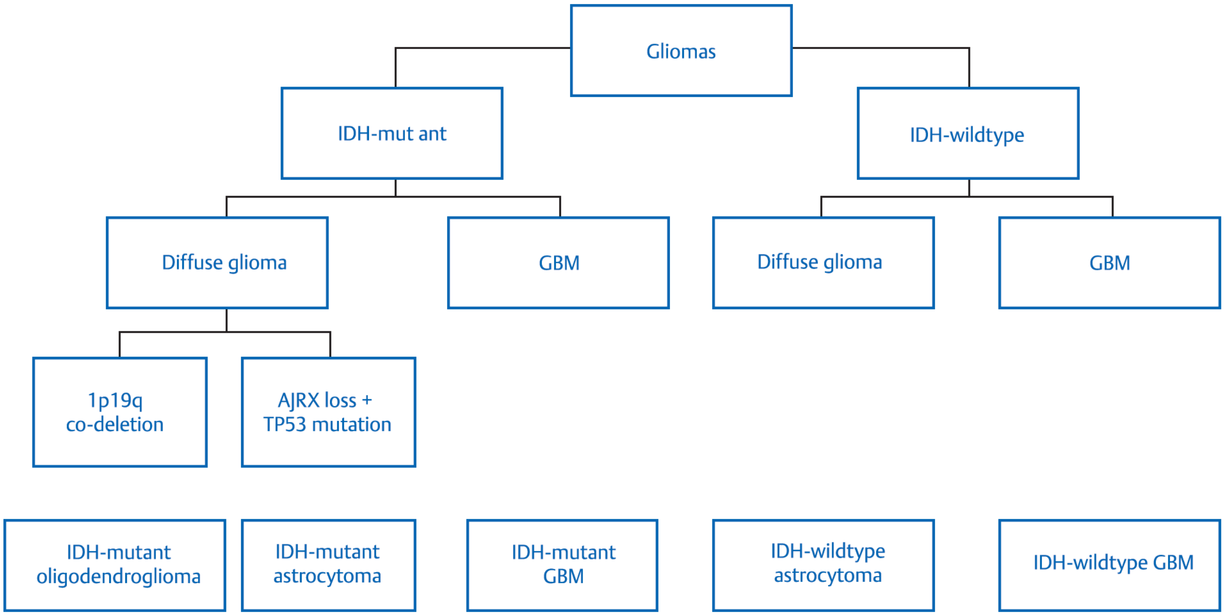
- No indication for steroid treatment or antiepileptic prophylaxis in the meantime.[8]

You advise her to proceed with surgical resection. The surgery and postoperative course were unremarkable. The final pathology report sates the following:

a. Diagnosis: oligodendroglioma grade 2
b. IDH 1: mutated and 1p19q co-deleted
c. *ATRX* is present and the MGMT promotor is hypermethylated

6. *How will the surgery impact her prognosis?*
   - Minimizing the volume of residual cancer is an important factor in cancer control, that is, progression-free survival (PFS) and overall survival (OS).[2,9,10]
   - Patients who undergo gross total resection have PFS of 7 years in contrast to a subtotal resection which yields a PFS of 3.5 years.[5]
   - The 5-year survival rate for patients with gross total resection is 90 to 95%, while that of a subtotal resection is 70 to 76%.[9,11]
7. *Following a complete resection of oligodendroglioma grade 2, what is the next step?*
   - Regular follow-up with brain MRIs every 6 months to evaluate for tumor recurrence
   - No indication for adjuvant therapy at this stage
8. *What is an IDH mutation? What is its significance in gliomas?*
   - IDH mutation definition:
     - An IDH mutation is a mutation that occurs in the gene that encodes the enzyme IDH 1 or 2.
     - Wild-type IDH converts isocitrate to α-ketoglutarate.
     - An IDH mutation alters the function of the enzyme such that it produces a neoenzyme that converts α-ketoglutarate to R-2 hydroxyglutarate (2-HG). 2-HG is a metabolite that remodels the methylation landscape of the genome of such cancers resulting in a distinct phenotype known as the CpG island hypermethylator phenotype (CIMP).
     - IDH 1 mutations occur exclusively in codon 132 while IDH 2 mutations occur in codon 172.
   - The significance of IDH mutation:
     - IDH mutation characterizes a distinct entity of gliomas that appear as grade 2 gliomas and then progress with time to a higher grade giving rise to what is known as secondary glioblastoma, as opposed to IDH wild-type gliomas that arise primarily as glioblastomas.
     - Therefore, IDH-mutant gliomas carry a better prognosis with a median OS of 2.5 years in patients with IDH-mutant glioblastoma multiforme (GBM) versus 1.3 years in patients with IDH wild-type GBM.[12]
     - As for anaplastic astrocytoma, the median OS is 5.5 years in patients harboring the IDH mutation versus 1.5 years in patients who do not.[12]
     - In addition, the presence of IDH mutation predicts the presence of MGMT promoter methylation (84% of IDH-mutant grade 2 gliomas).
9. *What does hypermethylation of the MGMT promoter mean? What is its significance? Four years following complete resection, the surveillance MRI is shown in* ▶**Fig. 17.3**. *Interpret the MRI.*
   - Hypermethylation of the MGMT promoter definition:
     - The MGMT is a DNA repair enzyme that counter acts the effects of DNA alkylating/methylating chemotherapies by removing the alkyl/methyl groups.
     - When the DNA promoter of the MGMT gene is hypermethylated, the expression of that gene is reduced resulting in greater sensitivity to chemotherapies such as temozolomide.
   - The significance of the MGMT prompter hypermethylation:
     - Predictive of a better response to temozolomide: the median OS is 23.5 months in patients harboring the MGMT methylation versus 12.6 months in patients who do not.[13]
   - On T2-weighted images, there is a diffuse hyperintense tumor at the surgical site with mild mass effect.
   - The contrast-enhanced MRI does not show enhancement in or around the mass.
10. *What is the natural history of recurrent oligodendroglioma? What will you advise the patient?*
    - Recurrent oligodendroglioma if left untreated will lead to death.
    - Surgery for diagnostic and therapeutic purposes: Minimizing the volume of cancer improves OS and given the risk of malignant transformation, it is necessary to treat these tumors as soon as possible.
    - A wait-and-watch approach cannot be a valid option for the reasons described above.

    The cancer is completely resected. The pathologist diagnoses the tumor this time as a grade 3 oligodendroglioma. IDH 1 mutated and 1p19q co-deleted.
11. *What is her prognosis in light of her tumor progression to a higher grade? What is your management plan?*
    - The management plan consists of:
      - Combination of radiation therapy and chemotherapy


Gliomas
IDH-mut ant
IDH-wildtype
Diffuse glioma
GBM
Diffuse glioma
GBM
1p19q co-deletion
AJRX loss + TP53 mutation
IDH-mutant oligodendroglioma
IDH-mutant astrocytoma
IDH-mutant GBM
IDH-wildtype astrocytoma
IDH-wildtype GBM

- The oligodendroglioma of gliomas has to be established histologocally first period to genetic testing
- • if genetic testing is inconciative or not done then the tumor is diagnosed based solvely on the histopathological features and labelled as NOS(e.g. astrocytoma NOS)

**Fig. 17.2** Glioma mutations. (Data adapted from Louis DN, Ohgaki H, Wiestler OD, Cavenee WK (Eds). WHO Classification of Tumors of the Central Nervous System (Revised 4th edition). IARC: Lyon 2016.)

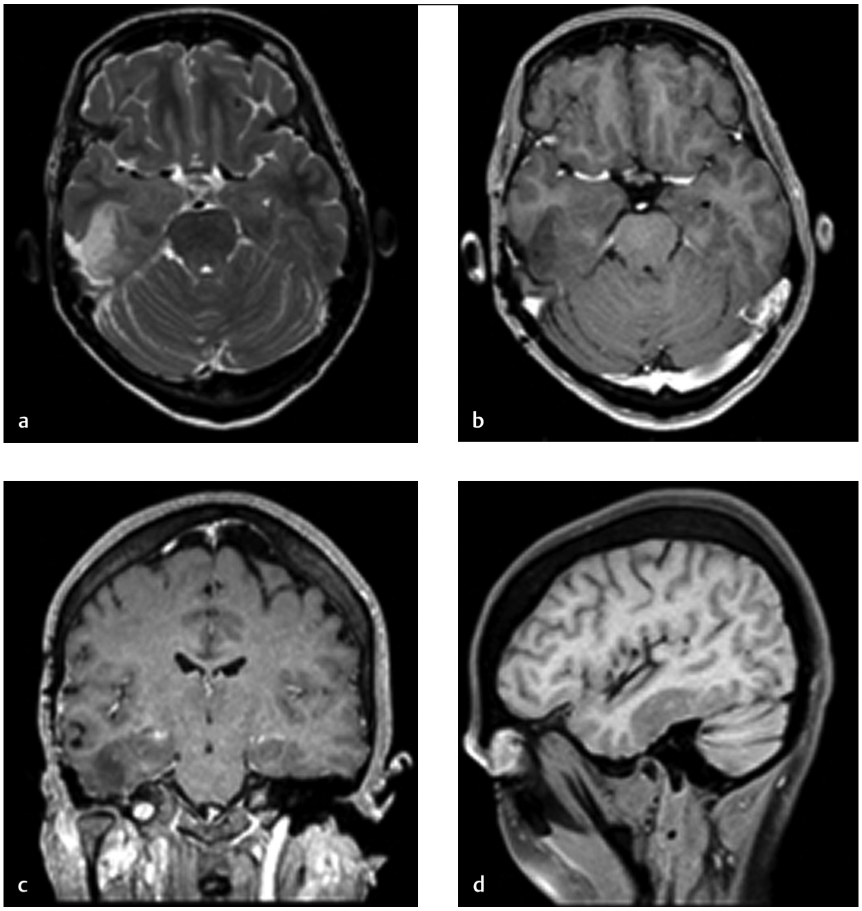


**Fig. 17.3** Brain MRI scan of patient described herein, with axial (**a**) T2- and (**b**) T1-weighed image, (**c**) axial and (**d**) sagittal T1-weighted image with contrast performed 4 years after surgical resection.

## Answers (*continued*)

- Chemotherapy can be procarbazine, lomustine, and vincristine (PCV) or temozolomide alone
- The prognosis[14]:
  - In patients with 1p/19q co-deletions, the median OS is 14.7 years in patients treated with PCV plus radiotherapy compared to 7.3 years in patients treated with radiotherapy alone.
  - In patients with non-co-deleted tumors, the median OS is 2.6 years regardless the treatment with PCV plus radiotherapy or radiotherapy alone.

12. *What are the key genetic alterations in primary and secondary glioblastoma?*

- Primary GBMs[11] have the following characteristics:
  - IDH wild-type
  - *TERT* promoter mutations (80%)
  - Homozygous deletion of *CDKN2A*/*COKN2B* (60%)
  - Loss of chromosomes 10p (50%) and 10q (70%)
  - *EGFR* alterations (55%)
  - *PTEN* mutations/deletion (40%)
- Secondary GBMs[11] have the following characteristics:
  - IDH mutant
  - *TP53* mutations (81%)
  - *ATRX* mutations (71%)
  - Loss of chromosome arm 10q

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# Case 18 Eloquent Cortex Low-Grade Glioma

*Ahmad I. Lary, Remi Nader, and Rolando Del Maestro*

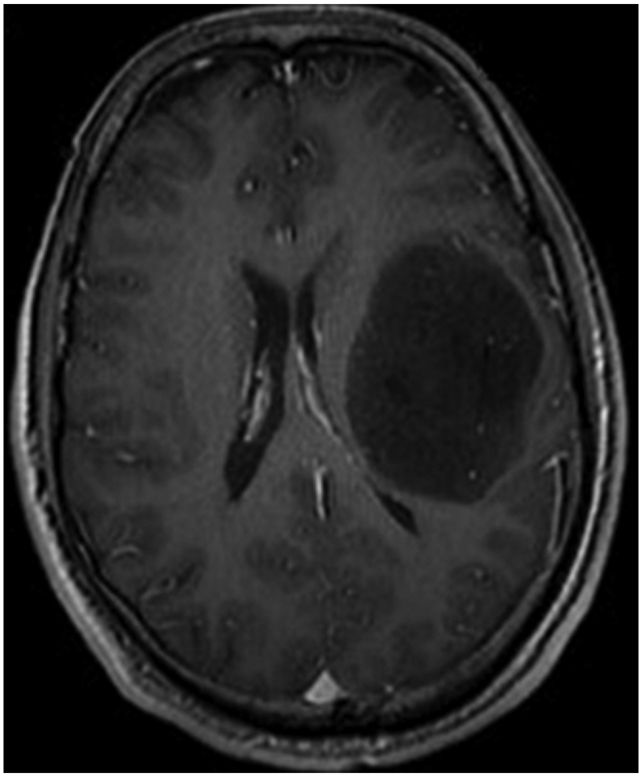

**Fig. 18.1** An axial T1-weighted magnetic resonance image.

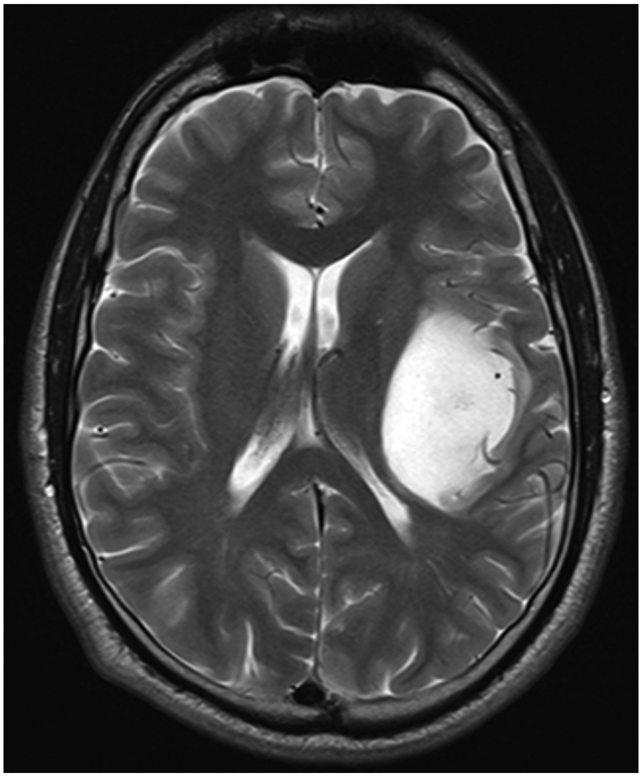

**Fig. 18.2** An axial T2-weighted magnetic resonance image.

## Clinical Presentation

- A 35-year-old male is referred from another hospital after suffering a single episode of a generalized tonic–clonic seizure.
- The patient was started on carbamazepine, 200 mg bid.
- He is right-handed and has no significant past medical history.
- Neurological examination demonstrated a mild facial weakness on the right side.
- MRIs were obtained and are shown in ▶**Fig. 18.1**, ▶**Fig. 18.2**, ▶**Fig. 18.3**, and ▶**Fig. 18.4**.

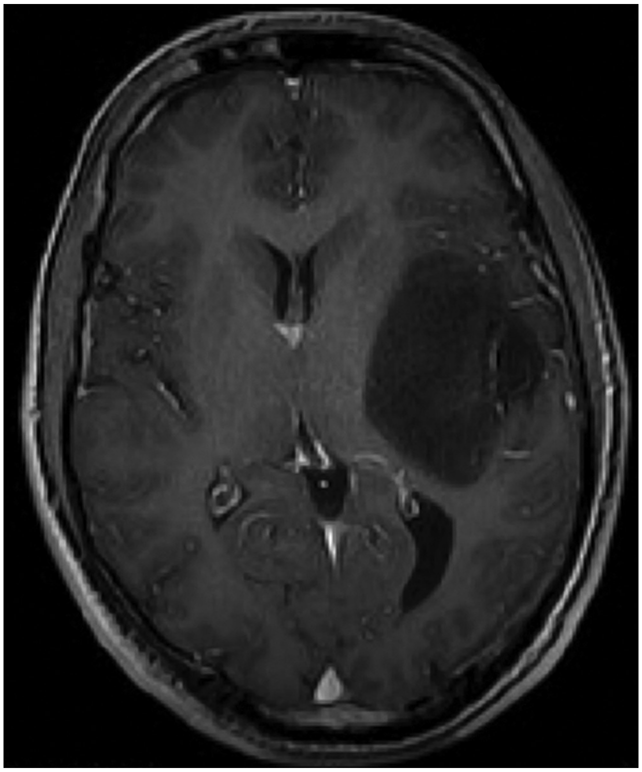

**Fig. 18.3** A sagittal T1-weighted magnetic resonance image with gadolinium.

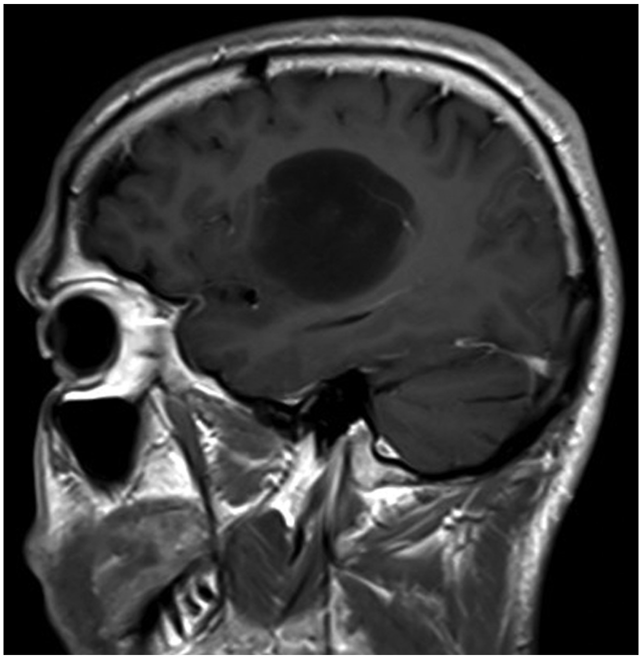

**Fig. 18.4** An axial T1-weighted magnetic resonance image with gadolinium.

## Questions

1. Describe the images.
2. What is the differential diagnosis?
3. Describe any further imaging investigations you would like to request.
4. You decided to resect this tumor, what potential complications are you most worried about that you will discuss in more detail with the patient?
5. Describe special intraoperative adjuncts you can utilize to aid you in the safe resection of this tumor.
6. What is the benefit of performing an awake craniotomy in this case?
7. Describe preoperative considerations in the planning of an awake craniotomy.
8. What measures can be employed to reduce the risk of permanent postoperative dysarthria/aphasia with dominant face motor cortex resection?
9. How is language mapping fundamentally different than motor mapping?
10. What are the common potential pitfalls of using too high of a stimulus during electrocorticography used for language mapping?
11. What measures should be considered if the patient sustains a seizure intraoperatively?
12. After the tumor was removed, the pathology showed "Oligoastrocytoma WHO II." Do mixed gliomas have a better or worse prognosis from pure astrocytomas, if yes then why?

## Answers

1. *Describe the images.*
   - There is a 5 × 4 × 4 cm intra-axial, insular mass on the left side.
   - The mass is hypointense on T1 and hyperintense on T2-weighted MRI.
   - There is a minimal mass effect with no significant midline shift.
   - There is no apparent surrounding edema.
   - The lesion is not enhancing on postcontrast images.
2. *What is the differential diagnosis?*[1]
   - Primary neoplastic: low-grade glioma (most likely diagnosis) (see prior case on glioma for a more elaborate differential diagnosis within primary neoplasms)
   - Primary neoplastic: primary central nervous system (CNS) lymphoma (these lesions are usually multiple and enhance with contrast)

## Answers (*continued*)

- Secondary neoplastic: metastases (these lesions are also usually multiple that enhances with contrast. They are typically seen in older patients or patients with other risk factors)
- Vascular: infarction (this diagnosis typically follows an arterial distribution)
- Infectious: cerebritis, herpes encephalitis (a common location of these lesions is the temporal lobe)
- Degenerative: demyelinating lesions (these lesions are usually multiple and periventricular in location)

3. *Describe any further imaging investigations you would like to request.*[2]
   - Functional MRI (fMRI): to demonstrate speech areas
   - Magnetic resonance spectroscopy (MRS): to help distinguish tumors from inflammatory or metabolic disorders[3]
   - Diffuse tensor imaging (DTI): to assess the white matter displacement by the lesion
   - One can also consider a CT scan to assess for calcifications and a vascular study (CTA or MRA) to evaluate vascular involvement and supply.
4. *You decided to resect this tumor, what potential complications are you most worried about that you will discuss in more detail with the patient?*
   - Speech disturbance (especially expressive dysphasia)
   - Weakness/hemiparesis along the right side of the body
   - Other common complications include seizures, hemorrhage, and infection
5. *Describe special intraoperative adjuncts that you can utilize to aid you in the safe resection of this tumor.*[2]
   - Intraoperative cortical mapping
   - Awake craniotomy
   - Neuronavigation
   - Intraoperative imaging: intraoperative ultrasound localization, intraoperative MRI, if available
   - Intraoperative microscope for magnification
6. *What is the benefit of performing an awake craniotomy in this case?*
   The involvement of left insula in this patient prompt the use of intraoperative insular awake mapping to obtain the best possible tumor removal with preservation of the eloquent structures (most importantly speech area in this patient).[4]
7. *Describe preoperative considerations in the planning of an awake craniotomy.*[5–10].
   - During pin fixation of the skull, local anesthetic with regional scalp block (1% lidocaine, 0.25% Marcaine, 1:200,000 epinephrine) should be used in conjunction with intravenous (IV) propofol for sedation.
   - A laryngeal mask airway (LMA) should be utilized during the initial portion of the procedure prior to patient awakening.
   - The dura should be infiltrated with lidocaine, especially in the region of the middle meningeal artery.
   - In order to avoid brain edema, the dura is opened after the patient is completely awake and cooperative.
8. *What measures can be employed to reduce the risk of permanent postoperative dysarthria/aphasia with dominant face motor cortex resection?*
   - Dominant facial motor cortex resections should be performed while continuously testing language.
   - The testing and dissection are started at the Sylvian fissure and slowly extended superiorly while avoiding the following structures:
     - Central artery
     - Descending motor fibers deep to the resection
     - Hand motor cortex superior to the resection
9. *How is language mapping fundamentally different than motor mapping?*
   - In motor mapping, a continuous current is used to provoke movement or sensation in an awake patient.
   - During language cortex mapping, the stimulation's purpose is to cause an electrical blockade of cortex function rather than eliciting function.
10. *What are the common potential pitfalls of using too high of a stimulus during electrocorticography used for language mapping?*
    - Provocation of a local seizure
    - Eliciting a false positive result (what is believed to be a language cortex is actually not) due to propagation of the signal to actual nearby language cortex
11. *What measures should be considered if the patient sustains a seizure intraoperatively?*
    - Adequate levels of antiepileptic medications should be secured perioperatively.
    - In order to arrest a seizure that is ongoing intraoperatively, one should consider the following:
      - IV short-acting benzodiazepine should be used such as diazepam or lorazepam.
      - Cold irrigation solution should be applied to the cortex.
    - Following a seizure, one should keep in mind that the mapping will be unreliable. One should, therefore, consider either an unmapped resection or abortion of the procedure: place a grid over the potential resection area of concern and prepare for extraoperative mapping with closure without completing the resection and reoperation at a later time.
12. *After the tumor was removed, the pathology came as "Oligoastrocytoma WHO II." Do mixed gliomas have a better or a worse prognosis from pure astrocytomas, if yes then why?*
    - A better prognosis, as the oligodendroglioma component has a better response to chemotherapy especially if it presents with LOH 1p19q.[11]

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# Case 19 Brain Metastasis

*Franz L. Ricklefs and Ennio Antonio Chiocca*

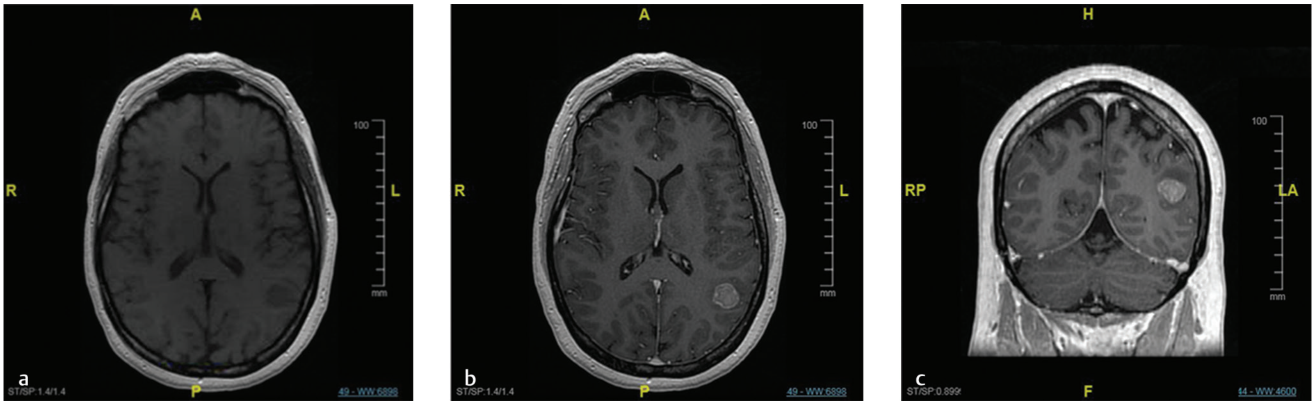

**Fig. 19.1** T1-weighted magnetic resonance image (MRI) of the brain without (**a**) and with contrast (**b, c**). Axial (**a, b**), coronal series (**a**).

## Clinical Presentation

- A 41-year-old woman presents to the emergency room (ER) with a generalized seizure during her work hours. She was unconscious for approximately 3 to 5 minutes. In her history, she reports being fatigued and having night sweats for the past 4 weeks.
- On neurologic examination, the patient is agitated but shows no neurological deficits.
- MRI is shown in ▶ **Fig. 19.1**.

## Questions

1. Interpret the MRI of the brain (▶ **Fig. 19.1**).
2. What is the initial work-up?
3. What is the differential diagnosis for a solitary enhancing mass lesion with cerebral edema? Name common primary sources for metastasis.
4. What is your initial management plan for this patient? How would you change it if the patient had an infratentorial lesion?
5. What are the treatment options for solitary and multiple brain metastasis? Describe the role for each modality.
6. Describe a treatment plan for this patient.

   The solitary mass was surgically removed. Histopathology revealed a melanoma metastasis. The patient received stereotactic radiosurgery (SRS) after surgery. Her positron emission tomography-computed tomography (PET-CT) showed several additional systemic metastases. Therefore, she received additional single-agent chemotherapy with dacarbazine.
7. What is the prognosis?
8. Discuss new treatment options, such as immunotherapy.

## Answers

1. *Interpret the MRI of the brain* (▶ **Fig. 19.1**).
   - There is a solitary 1.5 × 1.5 cm left parietal lobe mass with contrast enhancement.
   - There is surrounding edema but no mass effect.
2. *What is the initial work-up?*
   - Metastatic work-up includes:
     - A detailed general medical history: Is primary known? Has it been controlled for a long time?
     - CT scan of chest, abdomen, and pelvis
     - PET-CT
   - Brain MRI with contrast and spectroscopy
   - Total body PET scan
   - Laboratory testing including stool guaiac test and appropriate tumor markers

## Answers (*continued*)

3. *What is the differential diagnosis for a solitary enhancing mass lesion with cerebral edema? Name common primary sources for metastasis.*
   - Primary brain tumor (astrocytoma/glioblastoma multiforme)
   - Infection: bacterial, toxoplasmosis, *Cryptococcus* infection, aspergillosis, and herpes simplex encephalitis Inflammation: multiple sclerosis, tuberculosis gummas, granuloma, amyloidosis, sarcoidosis, and vasculitis
   - Others: resolving hematoma, lymphoma, infarction, and demyelination
   - Metastasis
     – Brain metastases from systemic cancer are the most common intracranial tumors and outnumber primary brain lesion roughly ten times.
     – Lung, breast, renal cell, colon cancer, and melanoma are the most common. Lung, colon, and renal cancers account for 80% of metastatic brain tumors in men.
     – On the other hand, breast, lung, colon, and melanoma cancers account for 80% of metastatic brain tumors in women.[1,2]
4. *What is your initial management plan for this patient? How would you change it if the patient had an infratentorial lesion?*
   - Patient should be admitted to the intensive care unit for close neurologic observation.
   - If the patient is highly symptomatic due to significant cerebral edema, the patient should be given steroids immediately: dexamethasone 10 mg intravenous (IV) bolus followed by 4 mg IV every 6 hours with gastrointestinal prophylaxis.
   - Consideration should be given to the use of prophylactic antiepileptic drugs as seizures occur in 20 to 40% of patients. However, the use of prophylactic antiepileptic medications is controversal.[3]
   - If an infratentorial lesion is present with compression of the fourth ventricle and acute hydrocephalus, surgical decompression needs to performed urgently as these signs can lead to devastating brainstem compression and potential herniation.[3]
5. *What are the treatment options for solitary and multiple brain metastasis? Describe the role for each modality*.
   - Current therapeutic options heavily depend on the clinical presentation of the patient and usually consist of a combination of surgery and radiation to control local and widespread disease progression.
   - The main treatment modalities for brain metastasis are surgery, whole-brain radiotherapy (WBRT), and SRS.
   - Several randomized controlled trials on patients with solitary brain metastasis that were randomly assigned to surgical removal of the brain tumor and WBRT or mainly WBRT[4–6] revealed a positive outcome benefit for the surgical group in terms of control of local and distant central nervous system (CNS) disease and death from neurologic causes. However, Mintz et al and a Cochrane review have challenged the benefit, since overall survival was not changed due to death from systemic disease.[7,8]
   - For multiple brain metastases surgical resection is favored if:
     – The metastases are surgically accessible
     – Immediate tumor debulking is needed
     – In cases of undiagnosed primary tumor
     – The lesion is causing hydrocephalus
     – In cases of radiation-resistant tumors (renal cell carcinoma, melanoma, and thyroid carcinoma)
   - Radiation alone is often used for patients with a poor Karnofsky Performance Score (KPS). Current regimes are either 20 Gy in 5 fractions or 30 Gy in 10 fractions. The combination of WBRT and surgery showed less frequent recurrence in solitary brain metastases[4] (18 vs. 70%) with a less likely cause of death due to a neurological reason (10 vs. 46%). For patients with two to four brain metastases, WBRT and surgery or radiosurgery improved the control of brain disease.[9] However, more recent studies are showing that there is no clear evidence of an effect on survival for WBRT and it is unclear whether WBRT may cause side effects such as memory loss.[10,11]
   - Stereotactic radiosurgery is beneficial for patients with inaccessible lesions or tumors with less than or equal to 3 cm diameter. Its further advantages are noninvasiveness, low radiation dose to normal brain tissue, and possible efficacy for radio-resistant tumors such as melanoma, renal cell carcinoma, and sarcoma.[12,13] For patients ≤ 50 years of age and with one to four brain metastasis, a survival benefit for SRS alone was shown in contrast to SRS + WBRT.[14] However, disadvantages of SRS include treatment restriction to lesions at or under 3 cm in greatest diameter, no mass reduction, prolonged use of steroids, and possible symptomatic radionecrosis.[15]
6. *Describe a treatment plan for this patient.*
   - Surgical intervention should be performed to obtain a histologic diagnosis and alleviate mass effect. Furthermore, it is critical to distinguish between an infectious process and neoplastic disease.
   - Radiation is usually required for metastatic tumors.
   - For infectious entities, the treatment options are radically different (i.e., IV antibiotics) and those can be tailored on the basis of microbiology and sensitivities to antimicrobials.
7. *What is the prognosis?*
   - Historically, the presence of a brain metastasis was correlated to a very poor prognosis. However, due

## Answers (*continued*)

to more aggressive treatment options and better systemic therapies, a slight improvement in overall patient survival has been achieved.

- If no treatment is undertaken, survival can be as low as 4 weeks.
- With the administration of high-dose glucocorticoids, survival increases to 8 weeks.
- WBRT can improve survival to 3 to 6 months.[4]
- For patients with a KPS of 70 or more, and no distant metastasis other than brain metastasis, a controlled primary focus and age less than 65 years (RPA class I) showed the best survival.[16]
- Patients with more than four brain metastases are usually not treated surgically and have a poor prognosis.[12]

8. *Discuss new treatment options, such as immunotherapy*.
   - Upcoming treatment regimens with immune checkpoint blockade are showing promising results with durable responses in metastatic tumors.
   - Anti-programmed cell death ligand 1 (PD-L1) antibodies are checkpoint inhibitors that interfere with PD-1 expressed on T cells, and its ligand PD-L1/-L2, expressed on tumor cells. Therefore, the T cells lose their inactivation resulting in an increased antitumor response. Two antibodies are currently available: nivolumab and pembrolizumab. Antibodies against PD-L1 are also becoming available.
   - Both agents have shown promising activity with durable responses in metastatic melanoma.[17,18]
   - There is an ongoing phase II study with pembrolizumab in patients with brain metastasis in non-small-cell lung cancer (NSCLC) and melanoma (NCT02085070, accessed March 2016). The drug is approved for advanced NSCLC.
   - It is unknown how the efficacy of these immune checkpoint inhibitors will affect the utilization and efficacy of surgery, SRS, and/or WBRT as treatment modalities.

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# Case 20 Meningeal Carcinomatosis

*Ramez Malak and Robert A. Moumdjian*

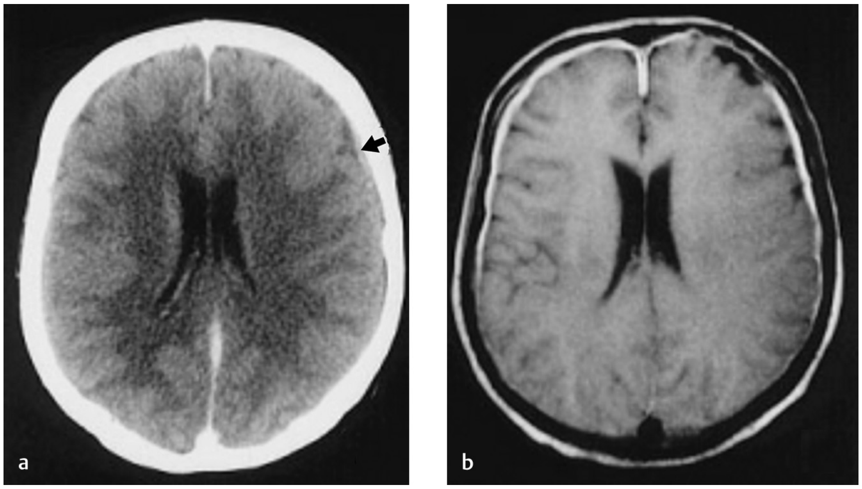

**Fig. 20.1** (**a**) Computed tomography scan of the brain and (**b**) T1-weighted axial magnetic resonance image of the brain, both with contrast. *Arrow* in (**a**) points to contrast enhancement of the meninges.

## Clinical Presentation

- A 45-year-old woman presents with headache, nuchal rigidity, diplopia, and weakness in the left leg.
- A CT scan shows contrast enhancement of the leptomeninges (▶**Fig. 20.1**).

## Questions

1. Provide a differential diagnosis of meningeal enhancement.
2. What are the most common causes of meningeal carcinomatosis (MC)?
3. What are the typical presenting symptoms?
4. What investigations would you obtain?
5. What is the yield of lumbar puncture (LP) in MC and how can you improve this yield?
6. What is the mechanism of invasion?
7. What are the characteristics of MC on craniospinal MRI?

   MRI of the lumbar spine (▶**Fig. 20.2**) shows multiple metastases along the cauda equina.

8. Describe symptomatic treatment.
9. Describe the advantage and complications of Ommaya reservoir placement.
10. Outline the treatment of MC.
11. How would you manage an accidental overdose of intraventricular methotrexate?
12. What is the prognosis of a patient with MC?

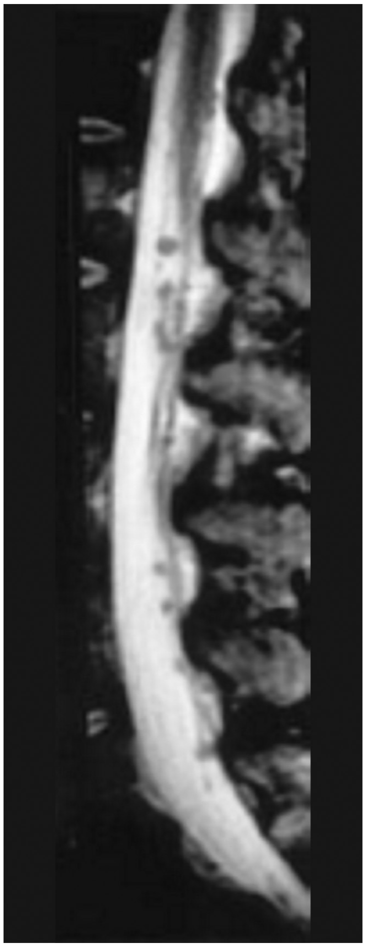

**Fig. 20.2** T2-weighted magnetic resonance image of the lumbar spine, midsagittal section.

## Answers

1. *Provide a differential diagnosis of meningeal enhancement.*
   - Neurogenic: increased cerebrospinal fluid (CSF) pressure, intracranial hypotension
   - Inflammatory: sarcoidosis
   - Infectious: subacute and chronic meningitis including tuberculosis, fungal infection, granulomatous cell infiltration
   - Vascular: local ischemia, venous thrombosis, hypoxia, subarachnoid hemorrhage
   - Traumatic
   - Drug induced: chemotherapeutic agents, heavy metals
   - Neoplastic: local tumor infiltration, primary meningeal glioma, primitive neuroectodermal tumor (PNET), isolated primary meningeal melanomas, rhabdomyosarcoma of the leptomeninges[1,2]
   - Other: ionizing radiation, metabolic disturbances, reaction to hyperventilation
2. *What are the most common causes of meningeal carcinomatosis (MC)?*
   - MC is estimated to occur in 5% of all patients with cancer and up to 20% in autopsy series[3]
   - MC is the third most common metastatic complication affecting the central nervous system (CNS) after brain metastases and epidural spinal cord compression, with 7000 to 9000 new cases diagnosed annually in the United States.[4]
   - There is a greater prevalence of solid tumors compared with hematologic malignancies.[5]
   - Solid malignancies presenting mostly with spinal or radicular symptoms:
     - Breast (12–35%) and lung cancers (10–26%)
     - Melanoma (5–25%)
     - Gastrointestinal cancer (4–41%)
     - Cancers of unknown primary (1–7%)
     - Head and neck cancers
   - Hematologic malignancies presenting more frequently with cranial nerve dysfunction or multifocal neurologic symptoms:
     - Leukemia
     - Lymphoma
     - Adenocarcinoma of unknown primary (ACUP)
3. *What are the typical presenting symptoms?*
   - Clinical manifestations that strongly suggest the diagnosis of MC include cauda equina symptoms or signs, communicating hydrocephalus, and cranial neuropathies.[4]
   - Contrary to bacterial or hemorrhagic meningitis, fever, photophobia, and meningismus are extremely uncommon.
   - The symptoms can be divided into two categories:
     - Hemispheric:
       - Headache, nausea, and vomiting
       - Alteration in mental status
       - Ataxia
     - Cranial nerve and spinal cord:
       - Diplopia (mostly cranial nerve VI)
       - Facial paresis
       - Lower motor weakness
       - Limb paresthesia
       - Back or neck pain and radiculopathic pain[3]
4. *What investigations would you obtain?*
   - CSF cytology
   - CSF biomarkers have been used but their utility is questionable because they have considerable variations in sensitivity and specificity[3]
     - Nonspecific biomarkers: β-glucuronidase, lactate dehydrogenase (LDH), beta 2-microglobulin, carcinoembryonic antigen
     - Organ-specific biomarkers: CA 15–3, CA 125, CA 19–9, CA724, AFP, NSE, Cyfra 21–1, and epidermal growth factor receptor (EGFR).
   - Radiologic studies
     - Cranial CT
     - Brain and spine MRI
     - Computerized tomographic myelography (rarely used)
   - Radionuclide CSF flow studies
     - Useful for the evaluation of CSF flow interruption due to tumor adhesions. Such interruption can lead to reduced treatment efficacy and increased toxicity due to impaired CSF drug distribution. CSF flow blocks can be efficiently treated with radiotherapy.[3,4]
     - Agents used for radionuclide study are:
       - 111Indium-diethylene-triamine pentaacetic acid
       - 99Tc macroaggregated albumin of choice
   - Meningeal biopsy from an enhancing region on MRI, in cases where CSF exams remain inconclusive
5. *What is the yield of lumbar puncture (LP) in MC and how can you improve this yield?*
   - The initial cytology is falsely negative in up to 40–50% of patients with pathologically proven leptomeningeal carcinomatosis. Diagnostic yield improves with:[2]
     - Repeated sampling (50% for the first to 90% for the third spinal tap)
     - CSF sample volume (10 cc at least)
     - Avoid delays in (i.e., immediate) processing and cytospin of the samples in the laboratory
     - Sampling site (LP provides a higher yield than ventricular CSF)
6. *What is the mechanism of invasion?*
   - Hematogenous spread via the arterial circulation or retrograde venous pathways along the valve-less Batson venous plexus
   - Perineural and perivascular lymphatics route

## Answers (*continued*)

- Direct spread from CNS tumors into the subarachnoid or ventricular spaces
- Iatrogenic spread during invasive procedures or neurosurgery through an ependymal or dural breach
- Common pathway: once malignant cells enter the CSF, they disseminate to distant parts of the CNS where they form secondary leptomeningeal deposits. The areas of predilection are: basilar cisterns, posterior fossa, and cauda equina[2]

7. *What are the characteristic aspects of meningeal carcinomatosis on craniospinal MRI?*
   - Enhancement and enlargement of cranial nerves
   - Superficial linear sulcal, cisternal, or dural enhancement
   - Irregular tentorial or ependymal enhancement
   - Cisternal or sulcal obliteration
   - Communicating hydrocephalus
   - Subarachnoid or intraventricular enhancing nodules
   - Multiple small nodular superficial brain nodules
   - Spinal linear enhancement
   - Spinal cord enlargement
   - Asymmetry of the roots with clumping of the roots of the cauda equina[1,2]
8. *Describe symptomatic treatment.*
   - Symptomatic treatment includes:
     - Pain relief using analgesics such as acetaminophen, opioids
     - Neuropathic pain often requires amitriptyline, clonazepam, or antiepileptic drugs (AEDs) such as Lyrica® (pregabalin) or Neurontin® (gabapentin)
     - Focal irradiation of symptomatic sites is often quite efficient in relieving pain
   - Seizures are managed with AEDs, but prophylactic administration of AEDs is not recommended in patients who have never had seizures.
   - Headaches related to edema or increased intracranial pressure can sometimes be treated with steroids.
   - In cases of hydrocephalus secondary to CSF blockade, a course of steroids during whole brain or skull base radiotherapy is sometimes appropriate, but shunting is often required.
   - Repeated LPs in the absence of cerebral edema or mass effect are often a good way to relieve headache.
   - Depression or fatigue may be managed with serotonin reuptake inhibitors or stimulant medication (methylphenidate).
   - End of life discussion is recommended in all patients.[3]
9. *Describe the advantage and complications of Ommaya reservoir placement.*
   - Advantages of Ommaya reservoir placement over repeated LP[6] are as follows:
     - Drug administration is painless and easier to perform
     - Better drug distribution in the entire subarachnoid ventricular space
     - Possibility of delivering frequent small doses of drug to reduce neurotoxicity
     - Can be used when the platelet count is around 20,000 cell/mm[3]
     - Provides a certainty that the drug has not been given in the epidural space (which is the cases in 10% of LPs)
     - Avoids some complications of LPs such as:
       - Local obstruction of CSF circulation secondary to arachnoiditis
       - Brain herniation in the presence of brain mass effect
     - When both a VPS and Ommaya ventricular access device are needed, an on–off valve may be placed
   - Complications[7]
     - If there is a concern due to risks for general anesthesia, it can be performed with local anesthesia.
     - Infections
     - Epilepsy by extravasation of the drug into the brain
     - Failure to puncture slit ventricles
     - Hemorrhages are rare complications occurring especially with repeated puncture attempts
     - Reservoir or catheter obstruction or dysfunction
10. *Outline the treatment of MC.*
    - The treatment of MC at present is palliative and rarely curative with a median patient survival of 2 months. However, palliative therapy often affords the patient protection from further neurological deterioration and consequently an improved neurologic quality of life.[5]
    - Some patients may be candidates for more aggressive treatment with radiotherapy and chemotherapy. Those patients should have the following criteria:
      - A good Karnofsky Performance Score
      - No evidence of bulky CNS disease by neuroimaging, because intra-CSF chemotherapy has a limited diffusion into tumor lesions with 2 mm or more diameter[3]
      - Absence of CSF flow block by radioisotope imaging
      - Expected survival more than 3 months
      - Limited extraneural metastatic disease[4]
    - Radiotherapy
      - Although it may stabilize or delay progression of neurologic symptoms, it does not prolong survival[5]
      - Radiotherapy is also indicated in the following instances:
        - Palliate symptoms, such as cauda equine syndrome and cranial neuropathies[4]

## Answers (*continued*)

        - To relieve CSF blocks which reduce the efficacy and increase the toxicity of intrathecal chemotherapy
    - Radiotherapy is administered at a dose of 30 Gy delivered in 10 fractions over 2 weeks. Common regimens are 20 to 30 Gy in 5 to 10 fractions to whole brain or to a partial spine field.[4] It provides effective relief of pain and stabilizes neurological symptoms but rarely leads to significant recovery.[2]
    - Irradiation of the entire neuraxis is too toxic in these patients who have generally already received multiagent chemotherapy and are prone to severe bone marrow toxicity.
    - Other experimental radiotherapy modalities include:
        - Tomotherapy and proton radiotherapy (with potentially less hematological toxicity)
        - Intra-CSF administration of radioisotopes or radiolabeled monoclonal antibodies[3]
- Chemotherapy
    - Chemotherapy can be administered intrathecally and/or systemically.
    - The majority of systemic chemotherapy does not penetrate the intact blood–brain barrier in adequate concentrations.[4] The goal of intra-CSF chemotherapy is therefore to bypass the blood–brain barrier, maximizing drug exposure in the CSF while reducing systemic toxicity.[3]
    - Methotrexate (MTX) is usually the first-line agent followed by cytarabine and Thiotepa.
    - MTX and cytarabine are used for leukemic meningitis whereas liposomal cytarabine and MTX for lymphomatous meningitis.[4]
    - Unfortunately, these drugs are not effective against many of the most frequent solid cancers associated with MC, particularly melanoma and lung cancer.[3]
    - Following the placement of an intraventricular catheter, MTX may be administered as 2 mg per day for 5 consecutive days every other week for four treatment cycles.[2–6] Equivalent volume of CSF (5–10 mL) should be removed prior chemotherapy administration.[3]
    - Neurologic complications of intra-CSF MTX include aseptic meningitis, acute encephalopathy, transverse myelopathy, and delayed leukoencephalopathy.
    - New regiments of chemotherapy for MC associated with solid cancer include:
        - Tyrosine kinase inhibitors (TKI) such as erlotinib for patients with non-small-cell lung cancer and sensitive EGFR mutations[3,8]
        - Capecitabine and hormonal therapy for patients with breast cancer
        - Immunotherapy agents such as ipilimumab for patients with melanoma[3]
    - Chemotherapy regiments under investigation are:
        - Temozolomide
        - Angiogenesis inhibitors (angiostatin) or vascular cell adhesion molecules[3]
- Prophylactic treatment in lymphoma and leukemia
    - Prevention of CNS relapse is increasingly the goal of primary therapy for patients with either non-Hodgkin's lymphoma (NHL) or acute lymphocytic leukemia (ALL). High-dose systemic and/or intrathecal chemotherapy are used depending on the presence of risk factors for CNS involvement.
    - These risk factors include:
        - Lymphoma grade and stage
        - Extent of extranodal disease
        - Young age
        - Elevated serum LDH levels
        - Presence of HIV-related NHL
        - Presence of a primary CNS lymphoma[5]

11. *How would you manage an accidental overdose of methotrexate?*
    - An accidental overdose of intra-CSF MTX is potentially fatal and can be managed with:
        - Immediate drainage of CSF via LP
        - Ventriculostomy with ventriculolumbar perfusion
        - Systemic steroids
        - Systemic leucovorin administration
        - Carboxypeptidase-G2 (CPDG2) (a potentially useful antidote)[3]
12. *What is the prognosis of a patient with MC?*
    - Despite therapy, median survival for meningeal carcinomatosis is about 4 months from diagnosis, and less than 15% of all patients survive 1 year following diagnosis. The median overall survival (OS) of untreated patients with leptomeningeal metastasis (LM) is 4 to 6 weeks.[3]
    - This is particularly true of patients with LM from solid tumors. Patients with breast cancer have a relatively better prognosis with median survival of 6 months from the time of diagnosis of MC.
    - Patients with leukemia and lymphoma have the best prognosis with MC. Such patients may respond rapidly and remain in a sustained remission for months to years.[6]
    - In addition to tumor histology, prognosis also depends on the performance status (PS), the age at LM diagnosis, and the treatment modality.[3]

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# Case 21 Primary Central Nervous System Lymphoma

*Hussam Abou-Al-Shaar, Randy L. Jensen, and William T. Couldwell*

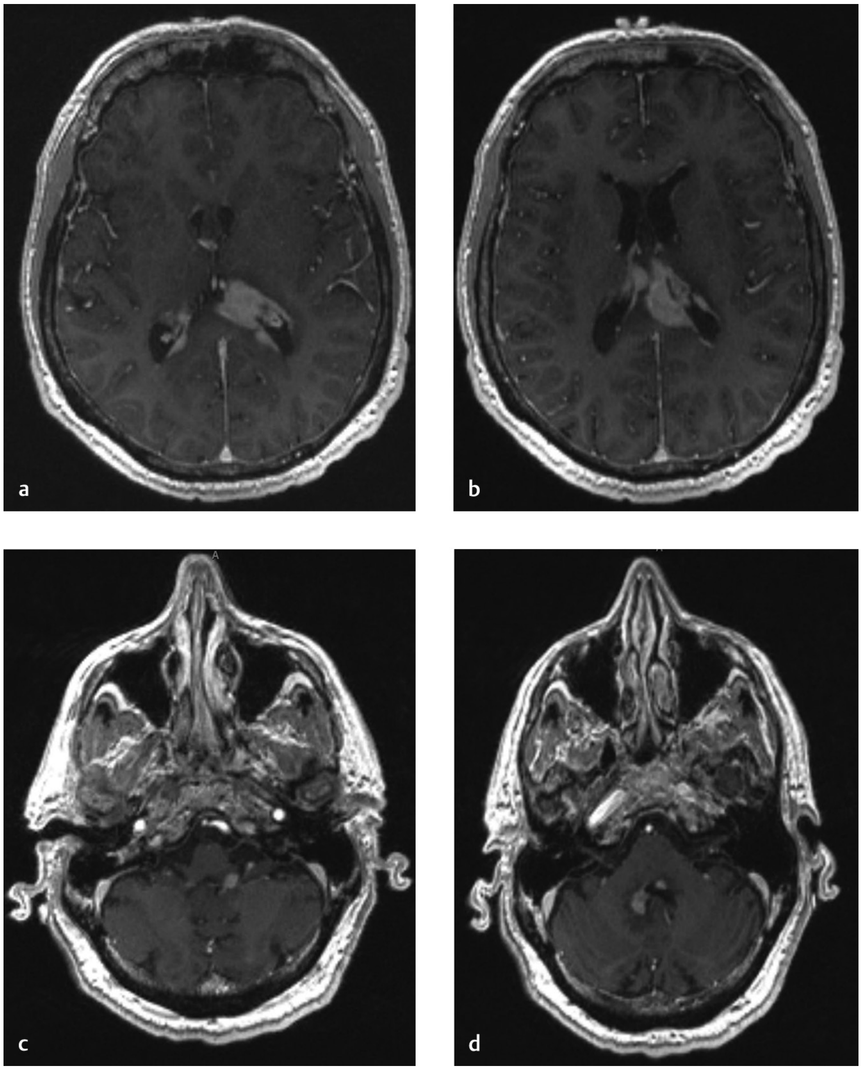

**Fig. 21.1** (**a–d**) T1-weighted magnetic resonance imaging of the brain with contrast depicting nodular enhancement throughout the ventricular system, with the largest focus at the atrium and occipital horn of the left lateral ventricle measuring 3.2 × 1.3 cm and a second focus along the atrium of the right lateral ventricle measuring 8.0 × 1.1 cm. There is hypointensity with mass effect and edema within the splenium of the corpus callosum and additional foci of nodular enhancement present within the frontal horn of the left lateral ventricles as well as within the fourth ventricle.

## Case Presentation

- A 71-year-old man presents with 1-week history of nausea, fatigue, and diplopia that has limited his ambulation.
- His past medical history is unremarkable except for hyperlipidemia and degenerative disc disease.
- He has no personal or family history of cancer.
- On examination, he was neurologically intact with no focal neurological deficits.
- CT scan showed multiple intracranial lesions and MRI of the brain is obtained (▶**Fig. 21.1**).

## Questions

1. What is primary central nervous system lymphoma (PCNSL)?
2. What is the epidemiology of PCNSL?
3. What is the appearance of PCNSL on CT and MRI?
4. What is the differential diagnosis of such lesions with imaging characteristics in immunocompetent patients?
5. What is the differential diagnosis of such lesions with imaging characteristics in immunocompromised patients?
6. What is the most common location for PCNSL?
7. Describe the histological appearance of PCNSL.
8. What is the treatment of PCNSL?
9. Describe the outcomes of patients with PCNSL.

## Answers

1. *What is primary central nervous system lymphoma (PCNSL)?*
   - PCNSL is an extranodal, malignant non-Hodgkin lymphoma that is confined to the brain, eyes, leptomeninges, or spinal cord, in the absence of systemic lymphoma.
   - Of all PCNSLs, 90 to 95% are diffuse large B-cell type, with the rest representing rare types including Burkitt (5%), lymphoblastic (5%), marginal zone (3%), or T-cell (2–3%) lymphoma type.[1]
2. *What is the epidemiology of PCNSL?*
   - It represents up to 1% of all lymphomas, 4 to 6% of all extranodal lymphomas, and about 3% of all central nervous system (CNS) tumors.[2,3]
   - It has a yearly incidence of 0 to 5 cases per 100,000 people, with a male-to-female ratio of 3:2.
   - Despite its relative rarity overall, PCNSL develops in 50% of patients with untreated acquired immunodeficiency syndrome (AIDS).
   - Among immunocompetent people, it most commonly occurs in men during their 7th decade of life, while in immunosuppressed patients, it tends to occur more in men during their 4th decade of life.[3]
3. *What is the appearance of PCNSL on CT and MRI?*
   - On CT scan, PCNSL is depicted as hyperdense avidly enhancing multifocal lesions.[4]
   - On MRI, PCNSL appears as an expansive, hypointense multifocal lesion in T1-weighted images and displays an avid homogeneous contrast enhancement with variable surrounding edema on contrasted T1-weighted images. Enhancement along the Virchow–Robin spaces is a highly specific feature of PCNSL.[5] It is isointense on T2-weighted images with restriction diffusion coefficient related to its high cellularity.
4. *What is the differential diagnosis of such lesions with imaging characteristics in immunocompetent patients?*
   - The differential diagnosis is broad and includes gliomas, metastases, ependymomas, abscesses, sarcoidosis, and multiple sclerosis.
5. *What is the differential diagnosis of such lesions with imaging characteristics in immunocompromised patients?*
   - The differential diagnosis includes various groups of pathologies, such as toxoplasmosis, tuberculoma, abscess, and progressive multifocal leukoencephalopathy.
6. *What is the most common location for PCNSL?*
   - In about 60% of patients PCNSL occurs as a single lesion.
   - The lesions are commonly found in the hemispheres (38%), thalamus/basal ganglia (16%), corpus callosum (14%), periventricular region (12%), cerebellum (9%), and isolated spinal cord (< 1%). Leptomeningeal lymphoma in the absence of a parenchymal mass represents < 5% of all PCNSLs.[1]
   - PCNSL tends to produce symptoms related to its location and mass effect.
   - PCNSL typically presents with neuropsychiatric symptoms as well as headache, confusion, lethargy, and focal neurological deficits; however, the occurrence of B symptoms (fever, weight loss, and night sweats) is rare.[6]
7. *Describe the histological appearance of PCNSL.*
   - PCNSL is a diffuse large B-cell lymphoma composed of large blastic cells often surrounding blood vessels, pleomorphic nuclei, and single or multiple distinct nucleoli. Tumor cells express pan-B cell markers, including CD19, CD20, CD79a, and MUM1. Tumor cells do not express proteins associated with plasma cell differentiation such as CD38 and CD138, but they may express CD10 and BCL2.[2]
8. *What is the treatment of PCNSL?*
   - The treatment of PCNSL is complex and grouped into different categories based on the patient's age and performance status.
   - In all cases, stereotactic biopsy should be the first step in the management of PCNSL patients to establish a definitive tissue diagnosis.
   - Steroid treatment should be avoided in stable patients before biopsy to avoid tumor shrinkage and misdiagnosis.[7]
   - Steroid treatment works rapidly to cause tumor shrinkage and decrease peritumoral edema. Despite its initial robust effect, most patients quickly relapse and require alternate treatment strategies.[7]

## Answers (*continued*)

- After establishing the diagnosis of PCNSL, a multimodal treatment composed of whole-brain radiation therapy and multiagent chemotherapy, with high-dose intravenous methotrexate (>3 g/$m^2$) as the backbone chemotherapeutic agent has been shown to prolong survival among both immunocompetent and immunocompromised patients.[8,9]
- Methotrexate, vincristine sulfate, and procarbazine hydrochloride followed by whole-brain radiation therapy and cytarabine in the post-radiation setting is one of the most commonly employed regimens in the management of PCNSL. It has shown an overall response rate of 91%, a progression-free survival of 24 months, and an overall survival of 36.9 months.[10]
- Among young patients, intrathecal methotrexate might be beneficial, especially in patients with concurrent leptomeningeal involvement.
- Among elderly patients, in whom chemotherapy is contraindicated, whole-brain radiation therapy with 40 to 50 Gy can be utilized. Unfortunately, this modality is not extremely effective with median survival rates of only 10 to 18 months. Furthermore, one of the feared complications of radiation therapy is delayed neurotoxic effects (i.e., subcortical dementia, gait ataxia, incontinence, as well as problems with attention, executive function, memory (particularly verbal), and psychomotor speed), especially in those older than 60 years.[11]
- Among human immunodeficiency virus (HIV)-negative immunocompromised patients, reduction of immunosuppression is usually beneficial to enable reconstitution of the immune system. Among HIV-positive patients with PCNSL, reconstitution of the immune system can be achieved with highly active antiretroviral therapy which has been shown to prolong survival. However, recent data have shown that both groups can be treated similar to immunocompetent patients with chemoradiotherapy.
- In general, surgery (gross total resection or debulking) for PCNSL does not improve survival. However, surgical resection may be mandated in certain cases such as when prompt reduction of intracranial pressure is necessary, for example, in patients showing herniation symptoms. More recent studies have also suggested that an attempt at gross total resection is reasonable for patients with solitary lesions that can be removed without morbidity; in these patients, survival may actually be prolonged when tumors are resected.[12]
- In addition, the international PCNSL Collaborative Group recommends staging work-up for all PCNSL patients to exclude systemic lymphoma and evaluate the extension of the disease. The staging work-up includes physical examination, brain MRI, contrast-enhanced CT of the chest, abdomen, and pelvis, fluorodeoxyglucose positron emission tomography (FDG-PET) scanning, cerebrospinal fluid (CSF) cytology and biochemical examination, bone marrow biopsy, ophthalmologic evaluation (including slit-lamp examination), and testicular ultrasonography (in elderly men because of frequent CNS involvement in testicular lymphomas).[13]

9. *Describe the outcomes of patients with PCNSL.*
   - The outcome remains unsatisfactory, with a survival of less than 20 to 30% at 5 years and a median survival of 10 to 20 months.[14,15]
   - The median survival of PCNSL patients without treatment is 3 months.
   - The complete remission rates of patients receiving both chemotherapy and radiation therapy ranges between 30 and 87%, and 5-year overall survival rates are 30 to 50%.[8,9]
   - Elderly patients tend to have poorer prognosis than young patients. Similarly, immunocompromised patients have a more dismal prognosis than immunocompetent patients.

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# Case 22 Fibrous Dysplasia of the Skull

*Burak Sade and Joung H. Lee*

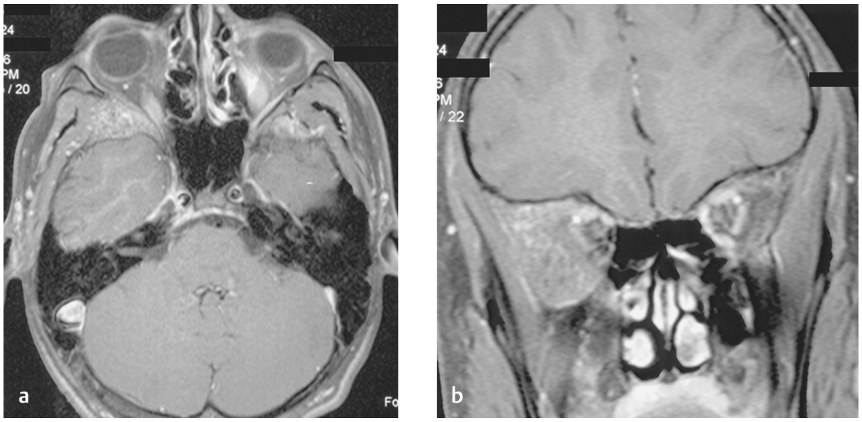

**Fig. 22.1** (**a**) T1-weighted postcontrast FatSat (fat saturation) axial, and (**b**) coronal magnetic resonance images. Note the hyperostotic abnormal bone involving the orbital roof, the posterolateral wall of the orbit, and extending into the infratemporal fossa.

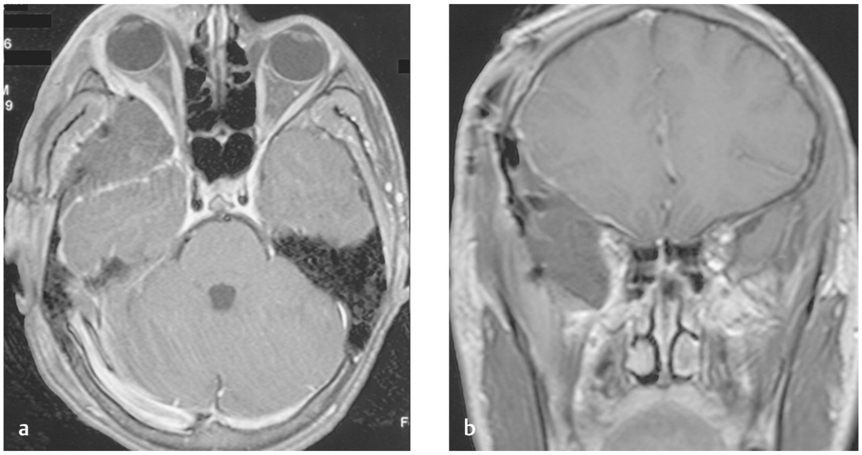

**Fig. 22.2** Postoperative T1-weighted postcontrast (**a**) axial and (**b**) coronal magnetic resonance images, confirming complete removal of the dysplastic bone.

## Clinical Presentation

- A 15-year-old right-handed boy is referred by a pediatric neurologist because of a right frontal, retro-orbital headache not responding to aggressive medical treatment.
- Neurologic evaluation is within normal limits.
- MRI scan of the brain is obtained (▶**Fig. 22.1**).

## Questions

1. Interpret the MRI scan.
2. What is your differential diagnosis?
3. What additional studies would you obtain?
4. What would be the indications for surgery based on your preoperative diagnosis?

Because of the intractable nature of his headaches, the patient was offered surgery. A right frontotemporal craniotomy was performed with a skull base approach consisting of extensive extradural removal of the lesion involving the orbital roof and the posterolateral wall of the orbit. The resection was further extended to the inferior orbital fissure inferiorly and foramen ovale posteriorly until normal bone texture was seen at all margins.

Postoperative MRI confirming complete resection of the tumor is shown in ▶**Fig. 22.2**.

The histology of the tumor was reported as “benign bone and fibrous tissue,” consistent with fibrous

## Questions (*continued*)

dysplasia. At 6-week follow-up, his headaches were significantly improved.

5. What would be your management goals?
6. What is the role of radiation therapy in the treatment of fibrous dysplasia?
7. What is the McCune–Albright syndrome?

## Answers

1. *Interpret the MRI scan.*
   - There is an enhancing hyperostotic lesion of the sphenoid bone on the right involving the orbital roof and posterolateral orbital wall.
   - It is extending into the infratemporal fossa to the level of the inferior orbital fissure.
2. *What is your differential diagnosis?*
   - The differential diagnosis includes:
     - Fibrous dysplasia
     - Fibro-osseous tumors (i.e., ossifying fibroma)
     - Primary bone tumors
     - Intradural process causing hyperostosis of the adjacent bone (i.e., sphenoid wing meningioma)
     - Metastasis
     - Other less likely diagnoses include lymphoma, rhabdomyosarcoma, Paget's disease, Langerhans cell histiocytosis, infection (osteomyelitis, soft-tissue infection, etc.)
3. *What additional studies would you obtain?*
   - CT scan: to better identify the osseous structures and assess the extent of the pathologic bone[1]
   - Neuro-ophthalmologic examination: because of the proximity of the lesion to the optic nerve[2]
   - Laboratory studies including serum alkaline phosphatase, calcium[3]
   - Possibly preoperative angiography or MR angiography with or without embolization, if surgery is planned
4. *What would be the indications for surgery based on your preoperative diagnosis?*
   - Indications for surgery include:[2,4]
     - Cosmetic concerns such as ocular proptosis or skull deformity
     - Compromise of the vision or ocular motility
     - Intractable headache or local pain
     - Rapid or aggressive growth
5. *What would be your management goals?*
   - Rather than being a true neoplasm, fibrous dysplasia is a result of the arrest of the bone maturation in the lamellar/woven stage.
   - Due to its polyostotic occurrence and poor demarcation from the normal bone, complete resection may be difficult at times or not possible. In one series, gross total resection was possible in only 25% of the patients.[5] Although subtotal resection may carry a risk for future regrowth of the residual lesion, its growth usually slows down after puberty.
   - Calcitonin or bisphosphonates have been used in multifocal or nonoperable cases.[2]
6. *What is the role of radiation therapy in the treatment of fibrous dysplasia?*
   - The role of radiotherapy is unproven.[4]
   - There may be a 44% of malignant transformation of fibrous dysplasia with radiation and therefore it is relatively contraindicated.[2]
7. *What is the McCune–Albright syndrome?*
   - The McCune–Albright syndrome is characterized by the following features:[2,6]
     - Fibrous dysplasia (usually polyostotic)
     - Precocious puberty and endocrinopathies
     - Areas of cutaneous pigmentation (café-au-lait spots)

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# Case 23 Orbital Tumor

Michel Lacroix

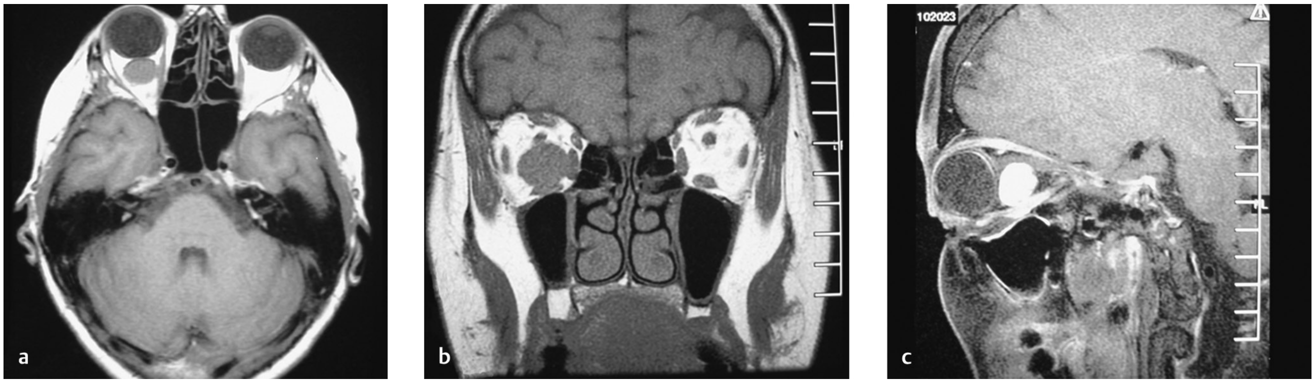

**Fig. 23.1** (**a**) Axial T1-weighted, (**b**) coronal T1-weighted, and (**c**) sagittal T1-weighted magnetic resonance images of the brain with gadolinium contrast brought by the patient during the initial visit.

## Clinical Presentation

- A 52-year-old woman presents with gradual proptosis.
- Examination reveals no diplopia, loss of vision, or other neurologic deficits.
- An MRI scan is shown in ▶**Fig. 23.1**.

## Questions

1. Interpret the MRI.
2. Give a general classification of orbital tumors.
3. What structures are contained in the annulus of Zinn?
4. Give a differential diagnosis.
5. What is your initial management?
6. Describe the surgical approach you consider best. What alternatives are available?

   You proceed with a lateral microsurgical approach, achieve a complete macroscopic resection, and the diagnosis is hemangioma.
7. What is the prognosis and what will be your follow-up?

## Answers

1. *Interpret the MRI.*
   - In the right orbit, there is a round lesion 1.3 cm in diameter. The lesion is homogeneous and is enhanced strongly after gadolinium injection.
   - The lesion lies inferiorly to the optic nerve to which it may or may not be attached, pushing it upward. There is a significant mass effect with displacement and/or invasion of the inferior rectus muscle and significant secondary proptosis.
2. *Give a general classification of orbital tumors.*
   - Based on their location and presentation with some specific symptoms, there are three categories of orbital tumors[1] excluding the primary ocular tumors. These are as follows:
     – Within the muscle cone (intraconal): visual loss and/or impaired orbital motility by mass effect; potential axial proptosis
     – Outside the muscle cone (extraconal): proptosis and orbital displacement; visual loss and impaired mobility by compression of individual muscles and deformity of the eye globe
     – Within the optic canal (intracanalicular): loss of vision and optic disc swelling; rare proptosis

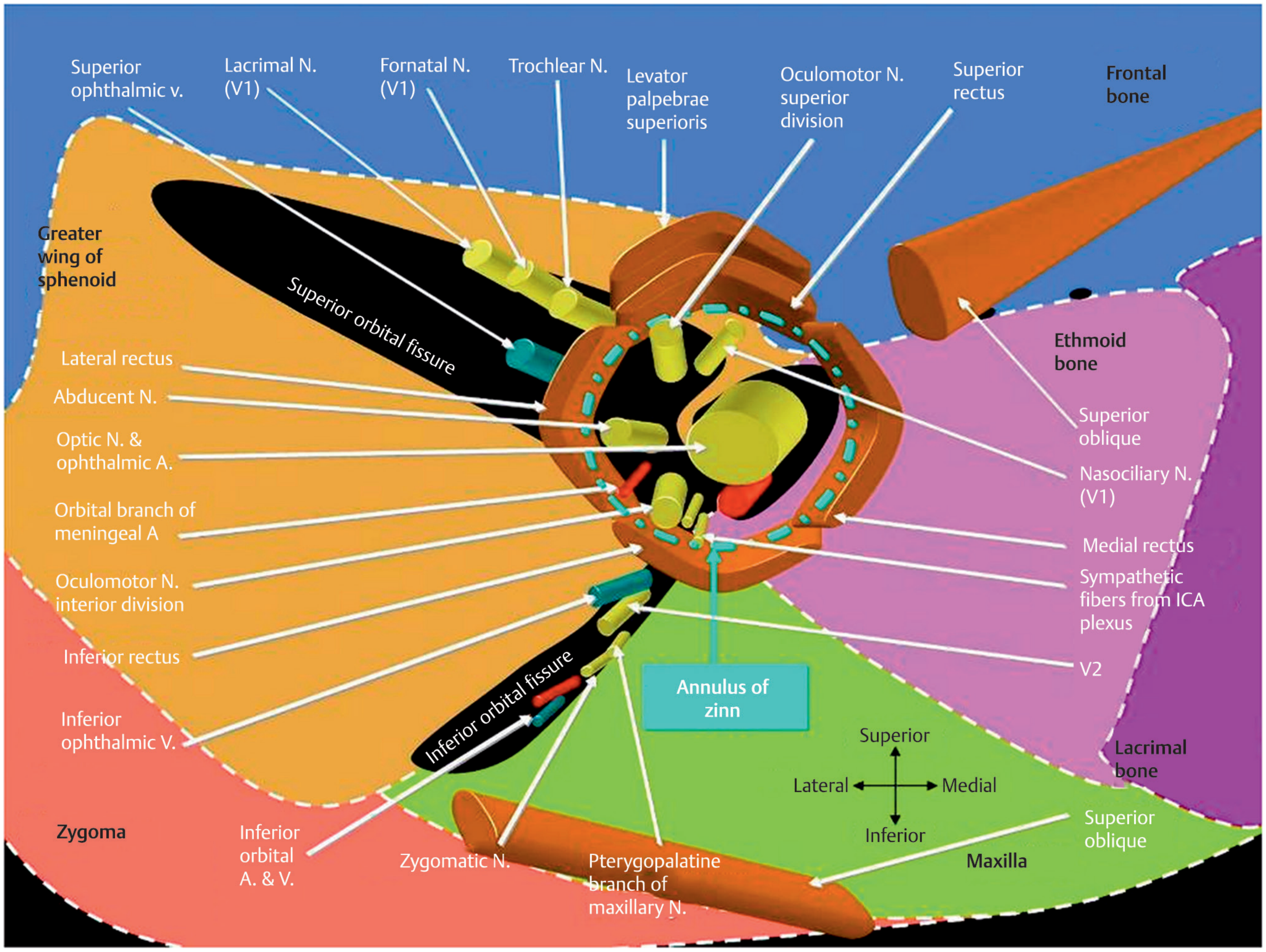
Superior ophthalmic v.
Lacrimal N. (V1)
Fornatal N. (V1)
Trochlear N.
Levator palpebrae superioris
Oculomotor N. superior division
Superior rectus
Frontal bone
Greater wing of sphenoid
Superior orbital fissure
Lateral rectus
Abducent N.
Optic N. & ophthalmic A.
Orbital branch of meningeal A
Oculomotor N. interior division
Inferior rectus
Inferior ophthalmic V.
Zygoma
Inferior orbital A. & V.
Zygomatic N.
Inferior orbital fissure
Pterygopalatine branch of maxillary N.
Annulus of zinn
Ethmoid bone
Superior oblique
Nasociliary N. (V1)
Medial rectus
Sympathetic fibers from ICA plexus
V2
Superior
Lateral
Medial
Inferior
Lacrimal bone
Superior oblique
Maxilla

**Fig. 23.2** Artist's rendering of the content of the annulus of Zinn.

## Answers (*continued*)

3. *What structures are contained in the annulus of Zinn?*
   - Cranial nerve (CN) II, ophthalmic artery, CN III (superior and inferior divisions), CN VI (nasociliary), CN VI (abducens)
   - See ▶**Fig. 23.2**
4. *Give a differential diagnosis.*
   - The lesion appears to be extra-axial and intraconal, and does not involve the eye globe. Considering the appearance of the lesion and the broad differential diagnosis of orbital lesions,[2] one should probably list these pathologies in the following order:
     – Hemangioma: the most common benign lesion of this location
     – Meningioma of the optic nerve sheath
     – Neurofibroma
     – Melanoma: the most frequent primary malignancy in adults in this location or other metastatic tumor
     – Lymphoma: a frequent cause of painless proptosis
     – Vascular, endocrine, infectious, and inflammatory diseases are unlikely. It is not a congenital malformation
5. *What is your initial management?*
   - Present all options to the patient including:
     – Observation/no treatment
     – Surgical resection for diagnosis and treatment
   - Present all the potential surgical complications including:
     – General (e.g., infection, hematoma, systemic complications)
     – Specific (e.g., loss of vision, diplopia, xerophthalmia, hypoesthesia)
   - Consult ophthalmologist for visual fields, extraocular movements' assessment, and surgical assistance.
   - CT scan is moderately helpful in this circumstance. The MRI provides all required imaging information. Consider selective angiogram.[3]
   - A metastatic work-up will not change the initial management.
6. *Describe the surgical approach you consider best. What alternatives are available?*
   - In general, anterior lesions are managed through a transorbital access, whereas apical lesions are

## Answers (*continued*)

managed with an extraorbital approach.[4,5] The algorithm of surgical access to the orbit suggested by Paluzzi et al[6] is insightful.

- Relying on the MRI, the lesion can be located inferior and lateral to the optic nerve and away from the optic apex. It is intraconal.
- A lateral orbitotomy is a viable surgical option:[7,8] a curvilinear incision is made from the superolateral aspect of the eyebrow, extended to the mid-lateral orbit, and is carried posteriorly ~3 cm. The temporalis muscle is reflected posteriorly.
- The lateral orbitotomy proper is performed after blunt dissection of the periorbita from the lateral epicanthus. The orbital rim is preserved for reconstruction. The orbitotomy is extended posteriorly and can reach the orbital apex if needed. A traction suture can be placed to identify the lateral rectus muscle and follows it. The incision in the periorbita is inferior to the lateral rectus muscle.
- A medial rotation of the globe is performed by light traction. Access is then achieved to the intraconal lesion.
- The tumor is dissected using a combination of cotton tip applicators and microinstruments and removed en bloc.
- A medial orbitotomy is preferred for a tumor located anteriorly to the orbit and medial to the optic nerve.
- Some posteriorly located medial lesions can be tackled via a combination of medial and lateral orbitotomies.
- For lesions with intracranial extension, i.e., lesions involving the optic canal and lesions medial to the optic apex, a transcranial fronto-orbital temporal approach is essential.
- Lesions located at the sphenoid wing and superior orbital fissure are best reached by a pterional approach.

7. *What is the prognosis and what will be your follow-up?*
   - A complete resection of a hemangioma is curative. No other treatment or long-term follow-up is required.9

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# Case 24 Multiple Ring-Enhancing Cerebral Lesions

*Roberto Rafael Herrera, José Luis Ledesma, Héctor P. Rojas, Kevin Petrecca, Rolando Del Maestro, and Francisco Sanz*

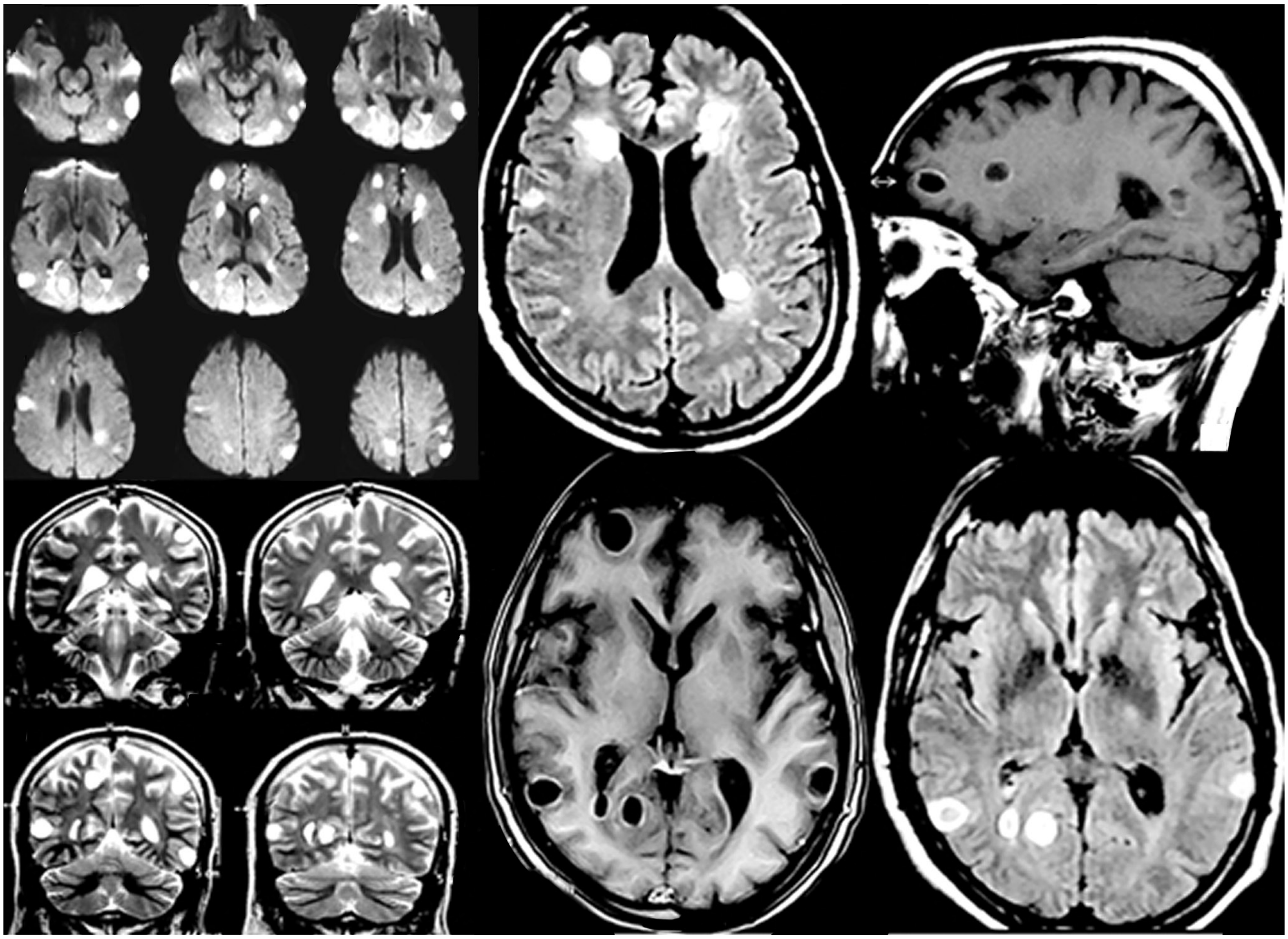

**Fig. 24.1** Magnetic resonance images with contrast, showing multiple intraparenchymal, supratentorial, and bilateral lesions (sequences T1, T2, fluid-attenuated inversion recovery, and gradient echo).

## Clinical Presentation

- A 55-year-old man presents with meningeal syndrome and depressed level of consciousness with a glasgow coma scale (GCS) score of 8/15. He is an engineer with a past medical history of hypertension on beta-blockers, obese, and ex-smoker.
- MRI obtained at admission of the brain is shown in ▶**Fig. 24.1**.
- Bacteriological cultures of cerebrospinal fluid, blood, and urine were performed. All results were normal.
- Empirical treatment was initiated with ceftriaxone and ampicillin. The patient improved clinically and after 7 days left the intensive care unit.
- On day 8, the patient experienced a sudden depression in his mental status with a GCS score of 4/15. Anisocoria with right mydriasis was observed. A brain CT scan was performed revealing extensive temporo-parieto-occipital right acute subdural hematoma (ASDH) with mass effect.

## Questions

1. Describe the MRI findings (▶**Fig. 24.1**).
2. What further information should be obtained from the clinical history?
3. What is the differential diagnosis?
4. What would be your initial investigations and how would you manage this patient?

## Questions (*continued*)

5. Are there any MRI findings that could distinguish a bacterial abscess from a fungal brain abscess or neoplasm?
6. What are the histologic stages of bacterial abscess formation and what are their radiological correlates?
7. What are the sources of bacterial abscesses?
8. What would be the next course of action if the patient deteriorated neurologically from an ASDH?

You elect to perform a surgical decompression and biopsy/resection. Cultures of one of these abscesses demonstrated methicillin-resistant coagulase-negative *Staphylococci* are sensitive to rifampicin, clindamycin, erythromycin, trimethoprim-sulfonamide (TMS), ciprofloxacin, vancomycin, metronidazole, and resistant to amoxicillin and cephalosporins first and second generation.

9. What is the next step in your management?

## Answers

1. *Describe the MRI findings (▸***Fig. 24.1***).*
   - At the supratentorial level, multiple nodular lesions are seen showing restriction on diffusion sequences; hyperintense signal on T2-weighted and fluid-attenuated inversion recovery (FLAIR) sequences are identified. The lesions described have a slight hyperintense halo on T1-weighted sequences and hypointense signals are identifiable on gradient echo (GRE) sequences.
   - These lesions are located along the cortical, subcortical, and periventricular areas in both cerebral hemispheres.
   - Limited perilesional edema is observed, not causing any mass effect.
2. *What further information should be obtained from the clinical history?*
   - It is important to identify risk factors for cerebral abscess formation and for metastatic disease, these include: smoking, weight loss, night sweats, fatigue, cough, intravenous drug use, cardiac or pulmonary abnormalities, human immunodeficiency virus status, other high-risk behaviors, etc.
3. *What is the differential diagnosis?*
   - The differential diagnosis includes multiple bacterial abscesses, neurocysticercosis, toxoplasmosis, tuberculomas, metastases, and multifocal glioblastoma.
   - By the morphology of the nodular lesions described, these could correspond to a central nervous system infection. The dual rim sign is not present in fungal abscesses, but a prominent peripheral rim or central susceptibility effects on susceptibility-weighted imaging (SWI; originally referred to as blood-oxygen-dependent [BOLD] venographic imaging) is seen. The appearance of pyogenic abscesses on SW MRI sequences depends on the used sequence, with the dual rim sign as a specific feature of pyogenic brain abscesses on SWI.[1]
4. *What would be your initial investigations and how would you manage this patient?*
   - Infection markers include an elevated white blood cell count, high erythrocyte sedimentation rate, and C-reactive protein
   - A chest X-rays to rule out pneumonia, tuberculosis, and neoplasm
   - Chest, abdominal, and pelvis CT for searching metastatic sites
   - Gram stain, blood and urine cultures to identify an hematogenous origin of the abscess as well as serum anti-toxoplasma titers
   - Western blot tests to detect *Taenia solium* antigens from serum
   - X-rays of the arms and legs to identify subcutaneous or muscular calcifications that can be present in cysticercosis
   - An electrocardiogram to assess cardiac rhythm and an abdominal ultrasound to identify any regional masses
   - It is important to have an accurate diagnosis; for this reason, a stereotactic biopsy of lesion located in a not eloquent area (and complete removal of the lesion if it is possible) should strongly be considered for histopathological studies.
5. *Are there any MRI findings that could distinguish a bacterial abscess from a fungal brain abscess or neoplasm?*
   - A conventional MRI study is useful to identify the lesions, determine their location and morphology, and allow a correct hypothesis as to their nature in most typical cases. However, the differential diagnosis from other brain lesions, such as nonpyogenic abscesses or necrotic tumors (high-grade gliomas and metastases), is often only possible through the use of functional sequences. These sequences include the measurement of diffusion with apparent diffusion coefficient (DWI-ADC), proton magnetic resonance spectroscopy (H-MRS), and perfusion weighted imaging (PWI), which complement the morphological sequences and provide essential information on structural, metabolic, and hemodynamic characteristics allowing greater neuroradiological confidence. Modern diagnostic MRI of pyogenic brain abscesses cannot be separated from knowledge, integration, and proper use of the morphological and functional sequences.[2]

## Answers (*continued*)

- SWI may be helpful in differentiating pyogenic abscesses from necrotic glioblastomas. The dual rim sign is the most specific imaging feature distinguishing the two and is typically seen in pyogenic abscesses.[3]
- A high-grade intralesional susceptibility signal (ILSS) may help distinguish glioblastomas from abscesses and necrotic metastatic brain tumors. The lack of ILSS or low-grade ILSS can be a more specific sign in the imaging diagnosis of abscesses.[4]

6. *What are the histologic stages of bacterial abscess formation and what are their radiologic correlates?*
   - There are four histological stages in cerebral abscess formation:[5,6]
     a. Early suppurative cerebritis (days 1–2): defined by endothelial cell swelling, perivascular neutrophil infiltration. CT findings include an area of hypodensity that may exhibit patchy enhancement.
     b. Late suppurative cerebritis with confluent central necrosis (days 3–7): defined by adjacent foci of necrosis which enlarge and become confluent. The infiltrate now includes macrophages, lymphocytes, and plasma cells. CT findings include a more pronounced central hypodensity with a thick enhancing ring surrounded by a hypodensity.
     c. Early encapsulation (days 8–14): defined by capsular neovascularity, fibroblast infiltration, collagen deposition, and perilesional edema. CT findings include a well-developed central core with a thinner well-formed enhancing ring.
     d. Late encapsulation (days >14): defined by central necrosis, a thin collagen capsule, and lymphocytes.
        Note: Capsule is thinner along the ventricular wall, increasing the susceptibility of rupture into ventricular system. CT findings include a very thin enhancing ring surrounded by a hypodensity.
7. *What are the sources of bacterial abscesses?*
   - There are following three sources of cerebral bacterial abscesses:
     a. The most common route is hematogenous which accounts for 25% of abscesses. The most common pathogen is *Streptococcus viridans*.
     b. A second etiology is from a contiguous source such as a paranasal sinus, middle ear, dental root, osteomyelitis, or emissary vein. The most common pathogen is *S. milleri*.
     c. The third route is direct from a trauma or post surgery, especially postsinus breach. The common pathogens are *Staphylococcus aureus* and *S. epidermidis.*
8. *What would be the next course of action if the patient deteriorated neurologically from an ASDH?*
   - As the etiology was not identified, the lesions were not decreasing in size even when broad-spectrum antibiotics were used and the patient is getting worse due to the hematoma; therefore, a more invasive approach is necessary. From the authors' point of view, a wide craniotomy along the right frontal lobe can be performed urgently to evacuate the ASDH and, in the same setting, one lesion close to the surface of the cortex can be resected for microbiology and histopathological studies.
   - Other less optimal or additional treatment options included antiepileptic medication to prevent seizures and steroid therapy for symptom relief.
   - After removing one or more lesions and obtaining the final etiologic diagnosis, appropriate treatment can be selected. Specific antibiotics according to sensitivities can then be initiated if the lesions are multiple abscesses. Alternatively, if the lesions are neoplastic, then treatment is tailored accordingly and may include chemotherapy, whole brain radiotherapy (WBRT), or radiosurgery for each lesion with or without WBRT.
9. *What is the next step in your management?*
   - The patient is diagnosed with multiple ring-enhancing brain abscesses produced by coagulase-negative *Staphylococcus*.
   - He should be treated with a combination of antibiotics, for example, vancomycin and metronidazole. Antibiotics should be continued for 6 weeks to 3 months (depending on infectious diseases service recommendation).
   - The patient may also need a peripherally inserted central catheter (PICC) line for long-term intravenous antimicrobial therapy and may also need home health therapy/nursing to monitor the PICC line.
   - Serial imaging and blood microbiologic and serum titers should be obtained to follow the progress of the treatment (including, complete blood count [CBC], erythrocyte sedimentation rate [ESR], and C-reactive protein [CRP]).
     He evolved with a very good outcome, recovering to close to his previous neurological status with a residual slight left hemiparesis and resolution of abscesses as seen on follow-up MRI studies (▶**Fig. 24.2**).

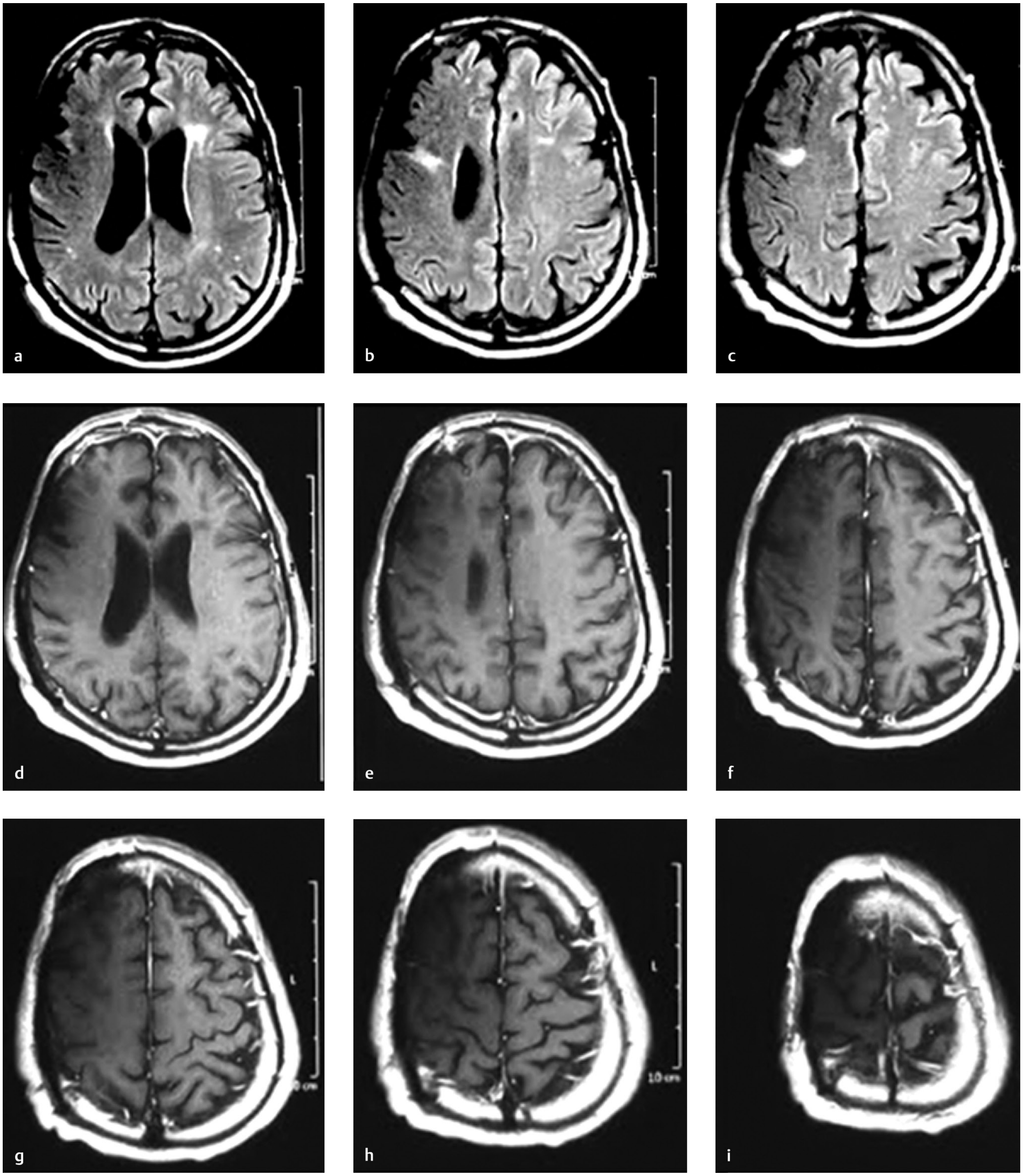

**Fig. 24.2** (**a–i**) Magnetic resonance images with contrast showing the evolution of multiple ring-enhancing brain abscess, treated with a combination of vancomycin and metronidazole.

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# Case 25 Paraganglioma

*Hussam Abou-Al-Shaar and Ossama Al-Mefty*

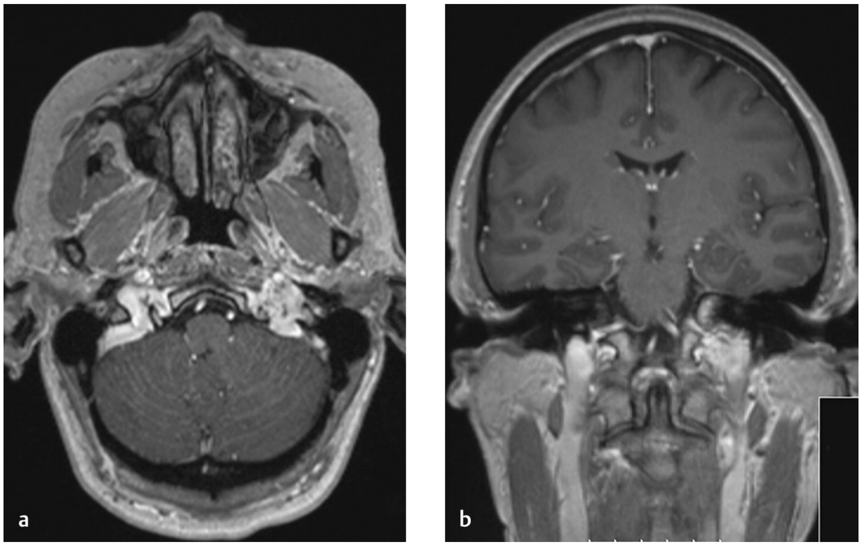

**Fig. 25.1** T1-weighted magnetic resonance image with contrast injection. (**a**) Axial cut taken at the level of the skull base and (**b**) coronal cut.

## Clinical Presentation

- A 44-year-old hypertensive woman with type 2 diabetes mellitus presents with a concussion. She had a history of speech difficulty and tongue wasting.
- The patient was noted to have ipsilateral tongue fasciculations, atrophy, and deviation. She also had slight ipsilateral sternocleidomastoid weakness with normal gag reflex and mild ipsilateral high-frequency sensorineural hearing loss.
- MRI scan with contrast is shown in ▶**Fig. 25.1**.

## Questions

1. Describe the MRI findings and explain what the CT scan would have shown.
2. What is the most probable diagnosis and what is the differential diagnosis?
3. What other studies would you order for this patient?
4. The serum epinephrine level test that you ordered was reported five times above normal level. What is the significance of this and what would you do next?
5. Interpret the angiogram (▶**Fig. 25.2**).
6. How would you describe the treatment options to the patient?
7. The patient chooses surgery. What preoperative and intraoperative measures would you take to avoid complications?
8. What would you do if the patient were also noted to have (a) an ipsilateral carotid body lesion or (b) a bilateral jugular fossa lesion?

   At the first follow-up visit, you discuss the pathology report with the patient which reads as follows, "… sustentacular cell density and the immunohistochemical staining with S-100 and chromogranin of the chief cells in this tumor was very low."
9. How would you interpret these pathologic findings?
10. The patient asks about the risk of her two sons getting this tumor. How would you address this issue with her?
11. Describe a classification system for glomus tympanicum tumors.
12. Describe the syndromes of the jugular foramen.

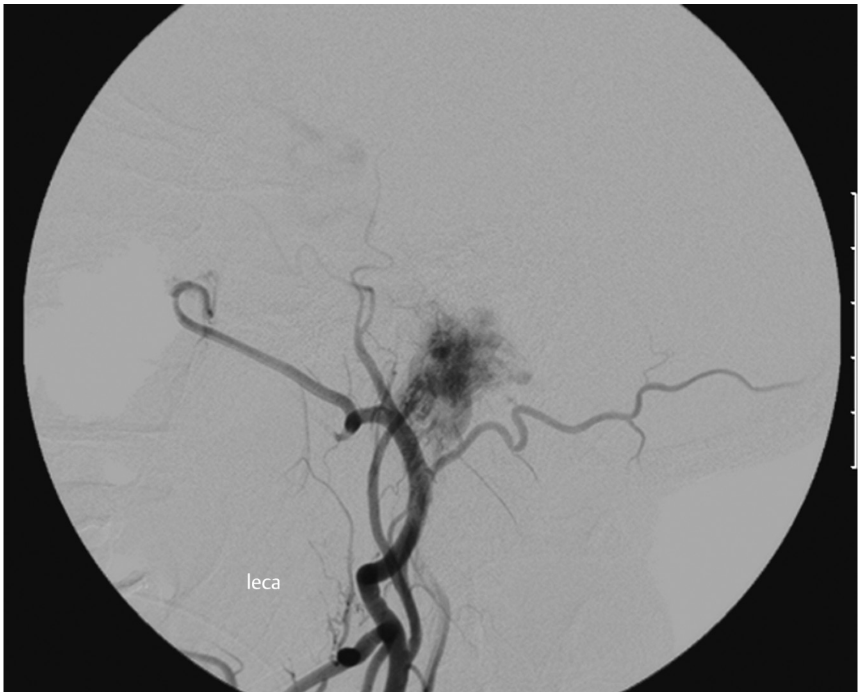

**Fig. 25.2** Left external carotid artery injection angiogram.

## Answers

1. *Describe the MRI findings and explain what the CT scan would have shown.*
    - T1-weighted MRI with gadolinium demonstrating a left-sided jugular foramen avidly enhancing lesion in the petrous bone noted to have "speckled pattern" suggestive of flow voids depicting high vascularity. Intracranial extension is not clearly evident in this section but could be expected.
    - A CT scan would have shown an eroded jugular foramen indicating the presence of a mass lesion in the petrous bone.
2. *What is the most probable diagnosis and what is the differential diagnosis?*
    - Tumors arising from the jugular foramen are most likely glomus jugulare (paraganglioma).
    - Other common possible lesions in the differential diagnosis include schwannoma and meningioma.
    - Other surrounding tumors may invade into the jugular foramen. These constitute chordoma, chondrosarcoma, giant cell tumor, cholesterol granuloma, endolymphatic sac tumor, temporal bone carcinomas, plasmacytoma, extension of nasopharyngeal carcinoma, metastases, etc.[1,2]
3. *What other studies would you order for this patient?*
    - Other relevant studies would include otoscopy that may show a red retrotympanic mass, angiogram, CT angiogram or MR angiogram, CT scan of the head, and CT scans of the chest, abdomen, and pelvis with or without $^{18}$F-fluoro-deoxyglucose positron emission tomography (FDG-PET). $^{123}$I-metaiodobenzylguanidine (MIBG) imaging might be considered instead of CT/PET to identify metastatic disease.[2,3]
    - Serum and urine metanephrine and catecholamine levels as well as urine vanillylmandelic acid levels
    - Glomus jugulare belongs to the neuroendocrine tumor family that includes other paragangliomas such as pheochromocytoma. Therefore, screening for a concomitant pheochromocytoma or another tumor, especially among genetically predisposed individuals is of paramount importance.[4] Moreover, screening such individuals for genetic mutations is important for detection of familial cases and for counselling.
4. *The serum epinephrine level test that you ordered was reported five times above normal level. What is the significance of this and what would you do next?*
    - Conversion of norepinephrine to epinephrine requires phenylethanolamine-*N*-methyl transferase (PNMT) that is present in the adrenal medulla. A very high serum epinephrine level raises the suspicion for pheochromocytoma. Glomus jugulare, when chemically active, generally produces norepinephrine.[5]
    - The patient should undergo α-blockade (e.g., phenoxybenzamine) prior to embolization or surgery. β-adrenergic blockade should be initiated if the patient develops tachycardia with α-blockade. Note: It is important *not* to initiate β-adrenergic blockade until α-blockage is fully established in order to avoid the possibility of unopposed α-adrenergic stimulation and hypertensive crisis.[4,6]
5. *Interpret the angiogram (▶**Fig. 25.2**).*
    - A left-sided external carotid angiogram demonstrating tumor blush fed via the ascending pharyngeal artery. Other arteries that may feed the glomus tumor are the occipital artery, internal maxillary artery, and the vertebrobasilar system.

## Answers (*continued*)

6. *How would you describe the treatment options to the patient?*
   - Surgery, radiation therapy (RT), radiosurgery (stereotactic radiosurgery [SRS]), combination of surgical debulking followed by RT/SRS, as well as observation have all been used for the treatment of glomus tumor.[3]
   - Patient age, comorbidities, tumor size and location, cranial nerve (CN) function, and hearing status are the important factors to consider while choosing the optimal treatment for the patient.
   - Indications for surgical resection include young age, a growing tumor, secreting tumors, presence of neurological deficits, significant intracranial mass effect, progression after failed prior treatment, malignant transformation, and low risk of lower CN injury.
   - RT is not tumoricidal and its effect is on the vascularity of the tumor. It is advocated for control, claiming a 20-year actuarial survival of 94% and a 20-year actuarial disease-free survival of 77%.[7]
   - The aim of radiosurgery is tumor control by delivering one fraction of 12.5 to 15 Gy. There has been no reported mortality and 2.1% recurrence rate, but the regression in tumor size is reported in a little less than one-third of the patients. It can be used for smaller recurrences (< 3 cm) and residuals. The results of surgical resection in the literature include early series where morbidity was higher than the current advanced techniques. In a meta-analysis of reported series, gross total resection was associated with a tumor control rate of approximately 86% and mortality rates of 3%.[8,9] However, with modern techniques that preserve the functions of lower CNs by intrabulbar dissection, a high rate of curative surgical resection with a marked reduction in morbidity in cases of complex and giant glomus tumors can be achieved.[10]
   - In one series, observation with serial imaging has been associated with stable or regressed tumor in 79% of the patients with new or worsening CN deficits in 30 to 33% patients during the period of observation. Another study showed that 52% tumors remained stable while 48% grew at a mean rate of 0.8 mm/year over a median follow-up period of 86.4 months.[11,12]
   - Therefore, close observation should be considered for elderly patients, those with significant comorbidities, and those who are asymptomatic. They can be observed with serial MRI every 6 months among symptomatic patients or every 12 months among asymptomatic patients.
7. *The patient chooses surgery. What preoperative and intraoperative measures would you take to avoid complications?*
   - As glomus tumor is a highly vascular tumor, preoperative embolization performed 1 to 2 days prior to the surgery prevents excessive bleeding in the operating room.
   - Preoperative embolization is recommended and has been shown to reduce the intraoperative blood loss and operative time with similar complication rate to non-embolized patients.[13]
   - Similarly, the patient should be started on α-, with or without β-blockers, for blood pressure control if the tumor is secreting catecholamines.
   - Intraoperative electroencephalogram as well as monitoring of CNs VII, IX, X, XI, and XII is recommended along with brainstem auditory evoked response and somatosensory evoked potentials.
   - The relationship of the CNs to the jugular foramen and hence, the tumor, are vital to the risk of damage to these nerves. The lower CNs at their exit are most vulnerable in the jugular foramen, where they run adherent to the outer wall of the jugular bulb. The described technique by Al-Mefty and Teixeira[10] in leaving the bulb inner wall at the foramen is critical in preserving these nerves function.[10]
8. *What would you do if the patient were also noted to have (a) an ipsilateral carotid body lesion or (b) a bilateral jugular fossa lesion?*
   - An ipsilateral carotid body tumor can be addressed in the same setting with the neck dissection that is performed with the tumor resection from the temporal bone.
   - If the patient has bilateral glomus tumors, as is seen in ~10% of sporadic cases and as high as 25 to 55% or more in familial cases, the resection of the opposite site should be performed only if the surgical resection on one side did not cause lower CN deficit.[10]
9. *How would you interpret these pathologic findings?*
   - Sustentacular cell density and the intensity of immunohistochemical staining of the chief and sustentacular cells are inversely proportional to the tumor aggressiveness.[14]
   - Anaplastic or metastasizing paragangliomas are either devoid or very depleted of sustentacular cells.[15]
10. *The patient asks about the risk of her two sons getting this tumor. How would you address this issue with her?*
    - Embryologically, glomus jugulare tumor is of neuroectodermal origin. The germline mutation is in succinate dehydrogenase subunits (*SDHD, SDHB, SDHC, SDHA*) of complex II of the mitochondrial respiratory chain transmitted in an autosomal dominant inheritance pattern with variable penetrance.[16]
    - *SDHD* is currently the leading cause of hereditary head and neck paraganglioma (> 50%), followed by *SDHB* (20–35%), and *SDHC* (15%) mutations.[16]
    - *SDHD* and *SDHAF2* gene mutations cause head and neck paraganglioma if they are inherited from the father only. If they are in an affected mother, as in this case, they get inactivated but may become active in the subsequent generations if one of her sons gets the gene in question.[5,16]

## ■ Answers (*continued*)

11. *Describe a classification system for glomus tympanicum tumors.*
    - There are two well-recognized classification systems that are summarized in ▶**Table 25.1**.[4]
12. *Describe the syndromes of the jugular foramen.*
    - Vernet syndrome affects CNs IX, X, and XI and occurs due to an intracranial lesion.
    - Collet–Sicard syndrome affects CNs IX, X, XI, and XII and usually occurs due to an extracranial lesion.
    - Villaret syndrome affects CNs IX, X, XI, XII, and the sympathetic nervous system and usually occurs due to a posterior retropharyngeal lesion.
    - Jackson syndrome affects CNs X, XI, and XII and usually occurs due to a vascular infarction of the medullary tegmentum.
    - Tapia syndrome affects CNs X, XII, and possibly IX and usually occurs due to a high cervical lesion.
    - See ▶**Fig. 25.3** for further illustration.[17,18]

**Table 25.1** Classification of glomus tympanicum tumors

| Type | Fisch Classification | Glasscock–Jackson Classification |
|---|---|---|
| A or I | Limited to middle ear cleft | Small; involves jugular bulb, middle ear, and mastoid |
| B or II | Limited to tympanomastoid without destruction of bone in the infralabyrinthine area | Extends to internal auditory canal with possible intracanalicular extension |
| C or III | Involvement of infralabyrinthine compartment and extending into the petrous apex along the carotid canal (subdivided into C1, C2, and C3) | Extends to petrous apex; possible intracranial extension |
| D or IV | Intracranial extension of tumor < 2 cm in diameter (D1) and > 2 cm in diameter (D2) | Beyond petrous apex to clivus and infratemporal fossa; possible intracranial extension |

*Source:* Adapted from Sampson and Wilkins 1996.[4]

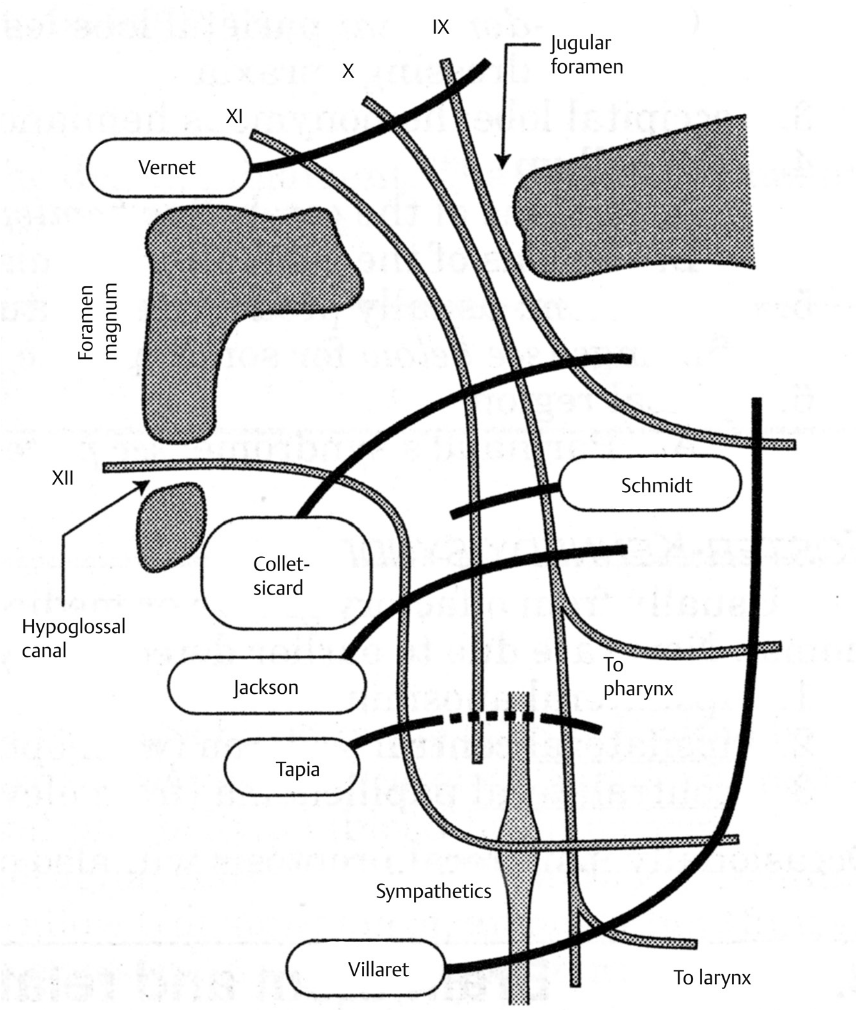

**Fig. 25.3** Schematic diagram of jugular foramen syndromes. (Reproduced from Greenberg MS. Handbook of Neurosurgery. 6th ed. New York: Thieme Medical Publishers; 2006:86)

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# Case 26 Colloid Cyst of the Third Ventricle

*Ahmed Alaqeel, Albert M. Isaacs, and Mark G. Hamilton*

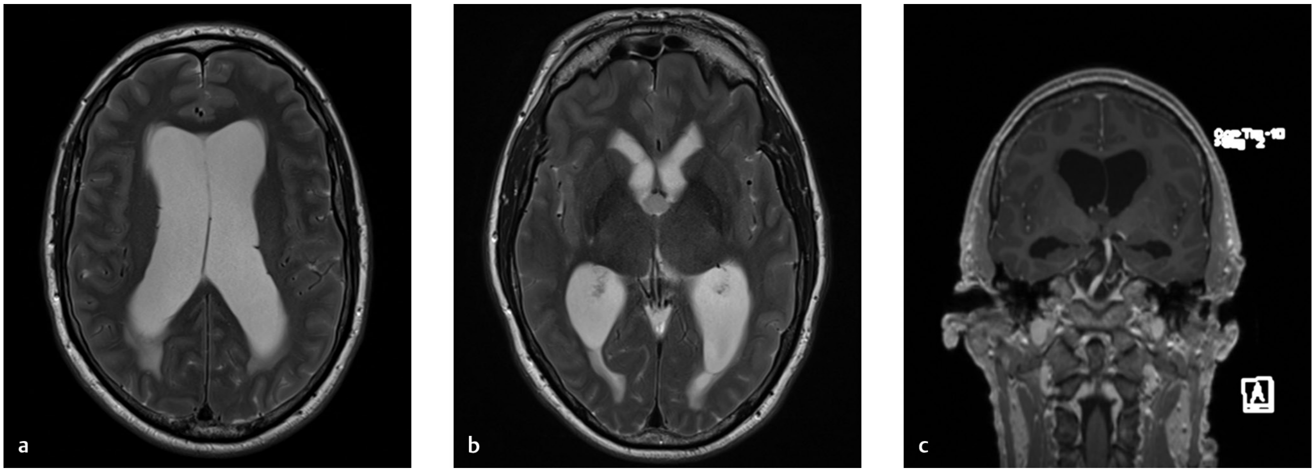

**Fig. 26.1** T2-weighted axial MR images (**a, b**), and a T1 weighted gadolinium enhanced coronal view (**c**).

## Case Presentation

- A 58-year-old male presents with 3-month history of severe headache, which was worse in the morning upon awakening and was associated with subjective mild gait difficulties.
- The neurologic examination was normal except for mild bilateral papilledema.
- He underwent a gadolinium-enhanced MRI of the brain (▶**Fig. 26.1**).

## Questions

1. Interpret the MRI scans.
2. Explain what a CT scan could demonstrate with this diagnosis.
3. What is the most probable diagnosis and what is the differential diagnosis?
4. What are the indications for surgery based upon the assumed diagnosis?
5. What are the surgical options?

Due to the concern about raised intracranial pressure, the patient underwent urgent endoscopic resection of the lesion. The lesion was confirmed to be a colloid cyst and a gross total resection of the lesion was achieved. An MRI done 3 months after surgery confirmed no residual colloid cyst and resolution of hydrocephalus as shown in ▶**Fig. 26.2**.

6. What are the advantages and disadvantages of endoscopic treatment versus microsurgical treatment of colloid cysts?
7. What is the incidence and natural history of the most likely diagnosis?
8. What would you recommend if the patient was asymptomatic?
9. How would you follow up this patient after colloid cyst resection?

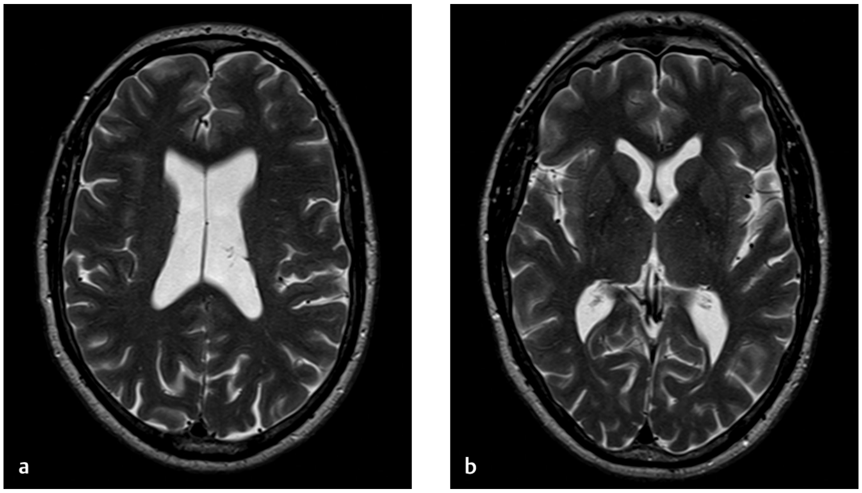

**Fig. 26.2** Postoperative T2-weighted axial MR images (**a, b**).

## Answers

1. *Interpret the MRI scans.*
   - The MRI scan shows a rounded cyst in the anterior third ventricle with obstructive hydrocephalus typical for a colloid cyst.
   - The T1-weighted and T2-weighted images demonstrate hyperintensity and isointensity, respectively, of the cyst contents.
   - There is a no significant peripheral or central enhancement. Small blood vessels are present on the lateral aspect of the cyst wall.
   - The MRI signal intensity of colloid cysts is notoriously variable, with any combination of T1 and T2 signal intensities described:[1]
     - The most common MRI appearance is hyperintensity with T1-weighted sequences and isointensity to hypointensity with T2-weighted sequences.
     - This variation is believed to be a result of the proteinaceous fluid as well as the paramagnetic effects of the metal ions in the fluid and hemorrhage.
2. *Explain what a CT scan could demonstrate with this diagnosis*.
   - CT scan would likely demonstrate an isodense, or calcified nonenhancing well-demarcated lesion within the roof of the third ventricle at the level of the foramen of Monro, with associated obstructive hydrocephalus.
3. *What is the most probable diagnosis and what is the differential diagnosis?*
   - The most probable diagnosis is colloid cyst of the third ventricle with obstructive hydrocephalus.
   - There is usually no significant differential diagnosis for a lesion of the third ventricle in this location with this MR appearance.
   - In cases with atypical imaging features, it is worth considering other masses which arise in the region of the foramen of Monro. These may include:
     - Calcified or hyperdense meningioma
     - Giant cell astrocytoma
     - Pilocytic astrocytoma
     - Central neurocytoma
     - Blood in the region of foramen of Monro
     - Aneurysm or abscess
     - Metastases
4. *What are the indications for surgery based upon the assumed diagnosis?*
   - Symptoms of raised intracranial pressure from hydrocephalus
   - Progressive ventricular enlargement meeting the definition of hydrocephalus, even in the absence of symptoms
5. *What are the surgical options?*
   - Urgent unilateral or bilateral ventricular cerebrospinal fluid (CSF) diversion is indicated in an obtunded patient to avoid cerebral herniation.
   - Current recommended surgical treatment options include:
     - Microsurgical resection (transcortical or transcallosal)
     - Endoscopic aspiration/resection
   - Stereotactic drainage has been tried in the past with poor long-term success.
6. *What are the advantages and disadvantages of endoscopic versus microsurgical treatment of colloid cysts?*
   - A recent meta-analysis of 1,278 patients evaluated aspects of both techniques.[2]
   - Open microsurgical technique was shown to have a higher rate of complete capsule resection and lower rate of reoperation as compared to endoscopic technique.
   - However, the complications of the open technique were significantly higher.
   - This study comes with the caveat that the timeline of data collection incorporates the development and maturation of the endoscopic technique, which unquestionably has undergone evolution and improvement over time.

## Answers (*continued*)

- Therefore, the advantage of open microsurgical technique is likely overestimated by this study, which is borne out by several case series of endoscopic technique that show resection and reoperation rates similar to that of open technique.[3,4,5]
- In sum, the operator should use the technique with which he is most familiar, and endoscopic technique is a safe and effective technical option when used by a skilled surgeon.[6]

7. *What is the incidence and natural history of the most likely diagnosis?*
   - The incidence of this lesion has been estimated at 3.2 per million per year, accounting for approximately 2% of all intracranial tumors.[7]
   - Patients with incidentally discovered colloid cysts could experience both lesion enlargement and symptom progression or, less commonly, contraction and symptom regression.
   - Incidental lesions rarely cause acute obstructive hydrocephalus or sudden neurological deterioration in the absence of antecedent trauma or spontaneous cyst hemorrhage.
   - Nearly one-half of patients with symptomatic colloid cysts present with obstructive hydrocephalus, which has an associated 3.1% risk of death.[8]
8. *What would you recommend if the patient was asymptomatic?*
   - Patients in whom asymptomatic colloid cysts are diagnosed can be cared for safely with observation and serial neuroimaging.[9]
   - Once lesions approach sizes greater than 1 cm, they are more likely to cause hydrocephalus, at which point an intervention is warranted.
   - Only a few centers treat asymptomatic colloid cysts without hydrocephalus[10] and this is not currently standard of care.
9. *How would you follow up this patient after colloid cyst resection?*
   - Colloid cysts can be cured after successful surgery and endoscopic colloid cyst resection results in a low overall recurrence rate.
   - MRI is the most effective modality available to follow patients for evidence of colloid cyst recurrence. Lack of recurrence by 10 years post resection is highly suggestive of surgical cure.

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# Case 27 Central Neurocytoma

*Turki Elarjani, Hussam Abou-Al-Shaar, Nazer Qureshi, Mohammad Almubaslat, and Abdulrahman J. Sabbagh*

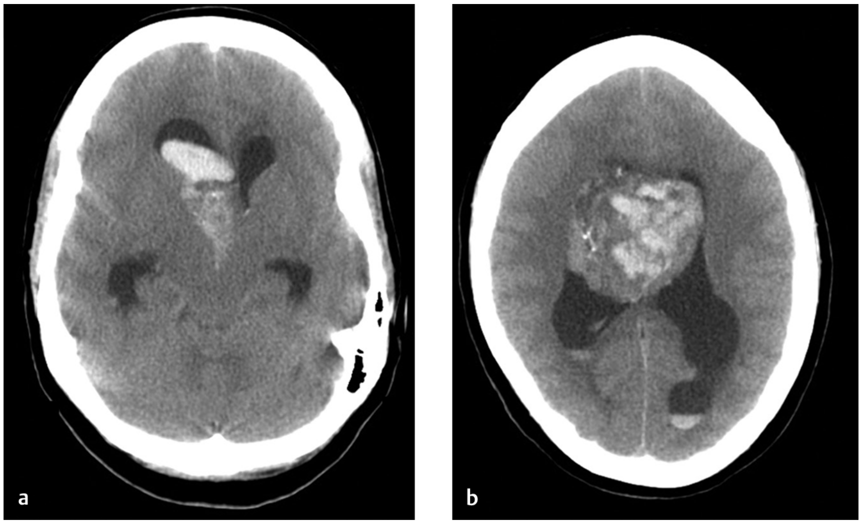

**Fig. 27.1** **(a, b)** Axial CT scan without contrast of the brain showing an intraventricular mass lesion.

## Clinical Presentation

- A 29-year-old woman pregnant woman presents to the emergency room (ER) with frequent episodes of tonic–clonic seizures.
- After undergoing a Cesarean section, she continued to have seizure episodes. She subsequently became unconscious with a decerebrate posture.
- An urgent CT scan of the brain was performed (▶ **Fig. 27.1**), at which time pupillary examination showed bilateral pupils reactive to light yet sluggish and unequal in size, the right pupil being 5 mm and the left pupil being 3 mm in size. Shortly thereafter, both pupils became fixed and dilated.

## Questions

1. Describe the findings demonstrated on the axial CT scan (▶ **Fig. 27.1**).
2. After delivery, the patient went into status epilepticus and was taken immediately for an urgent CT scan. After taking her out from the CT scan machine, you noticed that the patient has a fixed and dilated right pupil. What would be your next step in her care?
3. Describe the findings illustrated on the MRI scan (▶ **Fig. 27.2**).
4. What are your differential diagnoses?
5. Describe central neurocytoma. What is the prognosis of this lesion?
6. What are the different surgical approaches to this lesion? Describe them in terms of advantages, disadvantages, and complications.
7. The patient continues to have seizures despite being on antiepileptics, in addition to a decreased level of consciousness, and fever 1 week after the surgical procedure; cerebrospinal fluid (CSF) analysis reveals the following findings: CSF with 5,000 white blood cells per $mm^3$ (80% of neutrophils) and rare gram-positive cocci on direct microscopic examination. What is your diagnosis?
8. What are your interpretations of the MRI scan (▶ **Fig. 27.3**)? What are your treatment options?
9. What is the role of adjuvant therapies in a patient diagnosed with a central neurocytoma?
10. Describe ventriculitis. What is the prognosis of this condition?

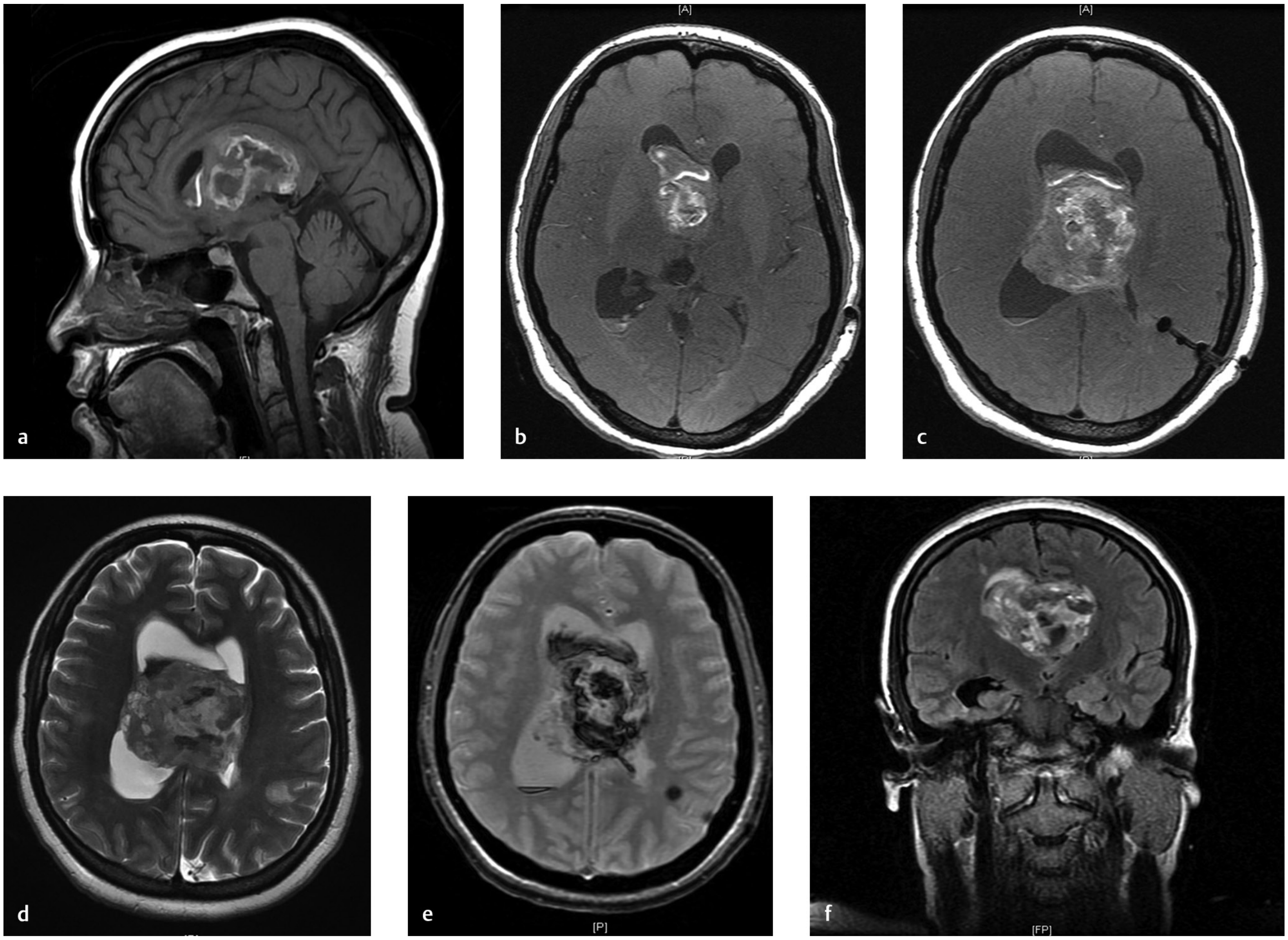

**Fig. 27.2** MRI scan of the brain; sagittal T1-weighted view with contrast (**a**); axial T1-weighted view with contrast (**b, c**), axial T2-weighted image (**d**); axial GRE image (**e**); coronal T2-weighted Fluid attenuated inversion recovery (FLAIR) sequences (**f**) showing a large intraventricular lesion.

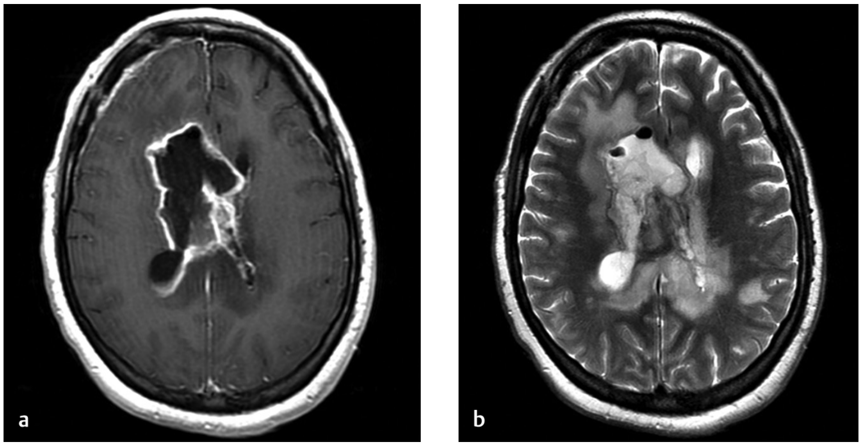

**Fig. 27.3** MRI scan of the brain, axial view T1-weighted with contrast (**a**) and T2-weighted image (**b**), demonstrating the postoperative result.

## Answers

1. *Describe the findings demonstrated on the axial CT scan* (▶**Fig. 27.1**).
   - Axial noncontrast CT scan of the brain showing an intraventricular mass lesion located mainly within the right lateral ventricle extending into the left lateral ventricle (▶**Fig. 27.1b**) and third ventricle through the foramen of monro (▶**Fig. 27.1a**).
   - The lesion size is approximately 6.5 × 5.5 cm. It is well circumscribed, lobulated, and heterogeneous in density. Hyperdense foci are appreciated owing to intralesional calcifications and hemorrhages.
   - Ventriculomegaly is appreciated in bilateral lateral ventricles, in addition to intraventricular hemorrhages within the occipital horns bilaterally with effacement of sulci due to elevated intracranial pressure.
2. *After delivery, the patient went into status epilepticus and was taken immediately for an urgent CT scan. After taking her out from the CT scan machine, you noticed that the patient has a fixed and dilated right pupil. What would be your next step in her care?*
   - Urgent external ventricular drain (EVD) insertion as a temporary measure to decrease intracranial pressure (ICP) and drain CSF, prior to microsurgical resection of the lesion; EVD is indicated in patients who have sudden rapid neurological status deterioration with evidence of hydrocephalus.
3. *Describe the findings illustrated on the MRI scan* (▶**Fig. 27.2**).
   - T1-weighted MRI with contrast (▶**Fig. 27.2 a–c**). demonstrating a heterogenously enhancing intraventricular mass. Axial T2-weighted MRI and coronal FLAIR sequence (▶**Fig. 27.2 d, f**) depicting the mass with hyperintense foci signifying calcifications, and hypointense areas signifying hemorrhage. Axial GRE sequence (▶**Fig. 27.2e**) showing large hypointense areas distributed within the mass, indicating hemorrhage and calcifications.
   - The mass is mainly located within the right lateral ventricle, abutting, but not traversing, the septum pellucidum as it grows toward the left lateral ventricle. There is an ipsilateral entrapped ventricle (ventriculomegaly), due to obstruction of the foramen of monro with more dilation of the right temporal horn, compared to left temporal horn.
   - Left parietal EVD tract can be identified (▶**Fig. 27.2 c–e**).
4. *What are your differential diagnoses?*[1]
   "CENTRAL MS" is a mnemonic used for intraventricular lesions:
   - C - Choroid plexus papilloma, choroid plexus carcinoma, colloid cyst, central neurocytoma (CNC), and cavernoma
   - E - Ependymoma, epidermoid, and dermoid
   - N - Neurocytoma
   - T - Teratoma and tuber
   - R - Rule out infection
   - A - Astrocytoma, arteriovenous malformation (AVM), aneurysm, and abscess
   - L - Lymphoma and lipoma
   - M - Meningioma and metastases
   - S - Subependymoma and subependymal giant cell astrocytoma (SEGA)
5. *Describe central neurocytoma. What is the prognosis of this lesion?*[1–3]
   - Rare benign (WHO II) intraventricular tumor commonly arising in the lateral ventricles; although less commonly extraventricular.
   - Comprises 0.1 to 0.5% of primary central nervous system tumors; mostly affects patients between the ages of 20 to 40 years. Males and females are affected equally.
   - Thought to originate from the septum pellucidum, ventricular wall, or fornix, arising from neuronal cells, neuronal progenitors, or multipotent precursor cells.
   - Presenting signs and symptoms are that of increased ICP such as headache, nausea, vomiting, papilledema, and visual deficits. Other presentations include seizures, memory disturbance, and motor weakness.
   - Grossly, central neurocytomas appear as gray friable lesions consisting of calcification and/or hemorrhage. Microscopically, they are inclined to have homogenous uniform cells with small, round to oval nuclei, and limited cytoplasm (perinuclear halo). Alternate areas of dense tumor cells and anuclear fine fibrillar matrix are commonly seen.
   - May be confused with oligodendroglioma microscopically, due to the close resemblance of cells architecture.
   - Overall, central neurocytoma has a good prognosis, with gross total resection achieving high progression-free survival rates. However, the effect on overall survival remains controversial.
6. *What are the different surgical approaches to this lesion? Describe them in terms of advantages, disadvantages, and complications.*[2,4,5,12,13]
   - Transcortical–transventricular route
     - Advantages:
       - ○ Most appropriate for tumors lateralized to one side
       - ○ Superior working space, especially if there is an associated ventriculomegaly
       - ○ Simple and flexible in traversing lateral ventricles
       - ○ Avoidance of damage to the fornix, parasagittal veins, and pericallosal arteries
     - Disadvantages/complications:
       - ○ Postoperative seizures (29–70%)
       - ○ Gerstmann syndrome if parietal approach
       - ○ Potential for cortical transgression

## Answers (*continued*)

- Transcallosal–transventricular route
  - Advantages:
    - Excellent for tumors extending to both lateral ventricles and third ventricle
    - Minimal brain retraction and injury
    - Major reduction in postoperative seizures (0–10%)
    - Continuous visualization of intraventricular anatomy
  - Disadvantages/complications:
    - Injury to bridging veins
    - Astereognosis and confabulation
    - Akinetic mutism
- General complications of surgery include:[6,7]
  - Venous infarction
    - Sacrifice of critical cortical draining veins
      - Avoid through preoperative planning and detailed review of imaging
    - Sagittal sinus thrombosis leading to venous infarction
      - Retraction injury: avoid excessive retraction, keep the surgical field moist
      - Injury during opening of the bone flap above the sinus
      - Overuse of coagulation in the region of the sinus
      - Hypercoagulable state of the patient, including dehydration
  - Bilateral cingulate gyrus retraction or thalamic injury
    - Results in transient mutism
    - Care must be taken when retracting deeper structures
    - Hemiparesis from injury to the motor cortex
    - Seizure
    - Arterial injury to pericallosal or callosomarginal arteries with infarction of the regions supplied by these vessels
    - Disconnection syndrome from wider opening of the corpus callosum
    - Internal cerebral vein injury via coagulation
    - Intraventricular hemorrhage from inadequate hemostasis
    - Other systemic problems: infection, myocardial infarction (MI), deep vein thrombosis (DVT), pulmonary embolism (PE), etc.

7. *The patient continues to have seizures despite being on antiepileptics, in addition to a decreased level of consciousness, and fever 1 week after the surgical procedure; cerebrospinal fluid (CSF) analysis reveals the following findings: CSF with 5,000 white blood cells per* $mm^3$ *(80% of neutrophils) and rare gram-positive cocci on direct microscopic examination. What is your diagnosis?*[8]
   - Bacterial ventri culitis
     - Nosocomial ventriculitis should be considered in patients who underwent an EVD insertion, in addition to signs and symptoms of infections, such as fever and abnormal CSF findings of CSF pleocytosis, decreasing CSF glucose with a reduced CSF glucose/serum glucose ratio.
8. *What are your interpretations of the MRI scan* (►**Fig. 27.3**)*? What are your treatment options?*[8,9]
   - Large intraventricular cavity suggestive of postoperative debulking of the tumor
   - Enhancing ependymal layer on T1-weighted MRI with contrast
   - To select the appropriate medication, factors such as the most likely implicated pathogen, patient's age, underlying comorbid conditions, and immune status must be taken into consideration.
   - Coagulase-negative and positive Staphylococcal infections are commonly encountered among the gram-positive organisms; resistant gram-negatives should be suspected in intensive care patients, including *Pseudomonas*, *Acinetobacter*, *Klebsiella*, and *Enterobacter* species.
   - Treatment should be promptly initiated with broad-spectrum antimicrobial medications encompassing both gram-positive and gram-negative organisms. Third-generation cephalosporins with rifampicin or fosfomycin should be started; intravenous and/or intrathecal vancomycin plus fourth-generation cephalosporins or meropenem should be administered in areas with high prevalence of methicillin-resistant *Staphylococcus aureus* (MRSA), methicillin-resistant *S. epidermidis* (MRSE), and multiresistant gram-negative rods.
   - Changes in the antimicrobial medications are performed according to the sensitivity and resistance pattern once the cultures are analyzed. Intravenous aminoglycosides should be administered in gram-negative bacterial ventriculitis if repeated CSF cultures show persistent presence of organism despite appropriate intravenous antibiotics.
   - Recommended duration of therapy is 10 to 14 days; however, if repeated CSF cultures show negative results, then a shorter duration of 5 to 7 days is appropriate with careful follow-up to confirm infection control. EVD replacement should be considered.
   - Fungal superinfection, predominantly *Candida* species, is a possible complication of prolonged broad-spectrum antimicrobial medication use; triazole medications, such as voriconazole or fluconazole with amphotericin B, are appropriate if fungal infection is established.
9. *What is the role of adjuvant therapies in a patient diagnosed with a central neurocytoma?*[2]
   - Radiation therapy (RT)
     - Recommended to prevent recurrences after incomplete resection, i.e., subtotal resection (STR)

## Answers (*continued*)

  - Major side effects include radiation necrosis
  - May be considered as a primary therapy if other treatment modalities are not available, or if the patient is not amendable to surgical resection
- Stereotactic radiosurgery (SRS)
  - Ideal option for the intraventricular location of CNC, as excess radiation is absorbed by CSF, rather than spilled to the adjacent parenchyma
  - Benefits of SRS over RT include its rapid dose fall-off and less treatment time
  - Promising treatment choice for residual and recurrent tumors
- Chemotherapy
  - Can be utilized for large tumors, where it is difficult to perform SRS
  - Can be used in select cases to avoid the long-term radiation side effects of RT and SRS
  - Multiple side effects depending on the combination used and overall limited role
  - Its role is primarily in cases of recurrent or progressive CNC (including anaplastic or atypical CNC).[10,11]

10. *Describe ventriculitis. What is the prognosis of this condition?*[8,9]
    - Bacterial ventriculitis is inflammation of the ventricular system as a result of a bacterial infection of the CSF. It is often associated with EVD insertion.
    - The incidence of EVD-associated ventriculitis ranges from 2—27%, with a mortality rate of 10-75%.
    - The most common implicated organisms are gram positive cocci (70% S. epidermidis and 10% S. aureus), while other organisms form the remaining 20% (15% formed by gram negative Bacilli such as E. coli, Klebsiella, and Pseudomonas; the remaining 5% formed by fungi such as Candida, and gram negative anaerobes).
    - Risk factors for EVD-associated ventriculitis include frequent EVD manipulation (e.g. multiple sampling and injections through the EVD catheter), prolonged EVD duration, and the presence of an associated intraventricular hemorrhage.
    - Attempts to prevent EVD-associated ventriculitis include the use of antibiotic impregnated ventricular catheter (which carries the risk of developing bacterial resistance), silver nanoparticle impregnated ventricular catheter, and prophylactic systemic antibiotics (not proven to decrease risk; may increase risk of bacterial resistance).
    - Nosocomial EVD-associated ventriculitis should be suspected in patient who have an EVD, in addition to the clinical signs and symptoms, such as high grade fever (reaching as high as 40 degrees Celsius), altered level of consciousness, headaches, nausea/vomiting, and meningism.
    - CSF diagnostic studies are used to identify the type and sensitivity of the culprit organism. However, EVD contamination and colonization may give a false positive results; thus, clinical manifestations should be taken into consideration, in addition to laboratory values.
    - CSF values usually show high protein content, low glucose content, pleocytosis, and positive gram stain and culture. Pro-calcitonin is an important marker to aid in diagnosis. Cell index calculation is valuable to reveal obscuring variables to CSF cell count, especially in the presence of intraventricular hemorrhage.
    - Ventrculitis is best depicted on T2-weighted images and DWI sequences as hyperintensity of the ependymal lining with diffsuion restriction, respecitvely. T1-weighted images with contrast also show contrast enhancement of the ependymal lining.
    - Once diagnosed, the EVD should be removed promptly and empiric antibiotics should be started. Treatment of Candida ventriculitis should be continued for 2 weeks after the last negative CSF culture.
    - The prognosis depends on the cultured organism, patient comorbidities, and prompt initiation of antimicrobial treatment.

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# Case 28 Clival Chordoma

*Hussam Abou-Al-Shaar and Gary L. Gallia*

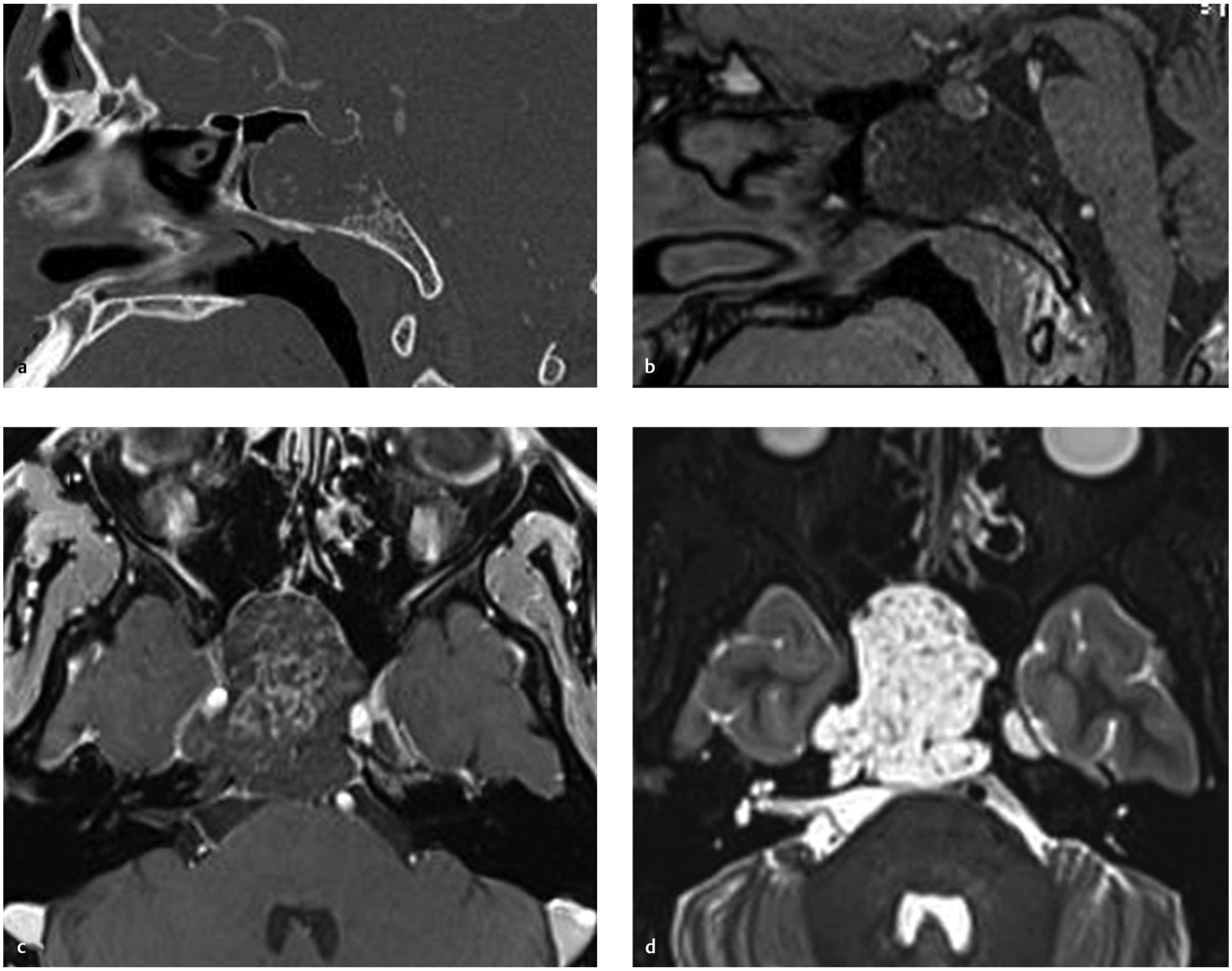

**Fig. 28.1** (**a**) Mid-sagittal CT scan with contrast demonstrates a destructive mass originating in the clivus with anterior extension into the sphenoid sinus. (**b**) Mid-sagittal T1-weighted MRI without contrast, (**c**) axial T1-weighted MRI with contrast, and (**d**) axial T2-weighted MRI demonstrate a T2 hyperintense heterogeneously enhancing mass centered in the clivus with posterior extension and mild mass effect on the pons as well as extension into the sphenoid sinus, right petrous apex, and right Meckel's cave.

## Clinical Presentation

- A 24-year-old man presents with several week's history of diplopia and headaches.
- On neuro-ophthalmological examination, he has an isolated sixth cranial nerve (CN) palsy in the right eye.
- CT and MRI scans are obtained (▶**Fig. 28.1**).

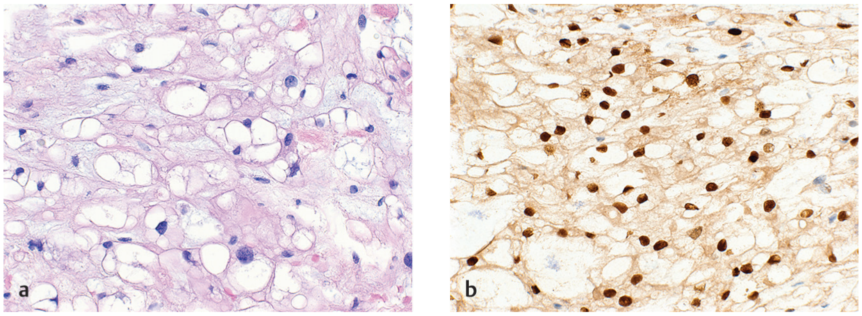

**Fig. 28.2** Histopathological findings in chordoma. Ovoid tumor cells are seen in a myxoid background. Diagnostic hallmarks include physaliphorous cells (**a**) and nuclear immunopositivity for brachyury (**b**).

## Questions

1. What is chordoma?
2. Describe the radiographic findings of clival chordoma (▶**Fig. 28.1**).
3. What is the proposed cell of origin for chordoma?
4. What is the epidemiology of chordoma?
5. What are the histopathological characteristics of chordoma (▶**Fig. 28.2**)?
6. What is the diagnostic marker of chordoma (▶**Fig. 28.2**)?
7. What are the common presenting signs and symptoms of clival chordoma?
8. What is the treatment of choice for patients with clival chordoma?
9. Discuss the various surgical approaches available to address this tumor.
10. What anticipatory measures should be undertaken when planning open surgery on this tumor?
11. What is the prognosis for patients with clival chordoma?
12. What is the role of chemotherapy in the management of patients with chordoma?

## Answers

1. *What is chordoma?*
   - Chordoma is a rare malignant tumor that occurs along the neuro-axis showing notochordal differentiation.[1]
2. *Describe the radiographic findings of clival chordoma (▶***Fig. 28.1***).*
   - On CT imaging, clival chordoma appears as a centrally located soft-tissue mass arising from the clivus associated with bony destruction. Calcification, contrast enhancement, and areas of low attenuation within the soft-tissue mass are other notable CT features of clival chordomas.[2,3]
   - On MRI, clival chordoma is iso-/hypointense on T1-weighted images and hyperintense on T2-weighted images. Heterogeneous enhancement is seen after gadolinium administration.[3]
3. *What is the proposed cell of origin for chordomas?*
   - Chordomas are thought to arise from transformed remnants of the primitive notochord.[4]
4. *What is the epidemiology of chordoma?*
   - Chordoma has an overall incidence of 0.08 cases per 100,000 individuals per year.[5,6]
   - Although there is some variability in smaller studies, there is a relatively even distribution between clival, spinal, and sacral sites of origin.[5]
   - The incidence of chordomas increases with age, with the median age at diagnosis in the sixth decade of life.[5,6]
   - Chordoma predominately occurs in Caucasians and has a slight male predominance.[5,6]
5. *What are the histopathological characteristics of chordoma?*
   - Macroscopically, chordomas are lobulated and exhibit a gelatinous matrix or more solid chondroid texture on cut section.[1]
   - There are three histological variants of chordomas:[1]
     - Chordoma, not otherwise specified (NOS), is the most commonly encountered histopathological variant. This subtype, which has also been referred to as classical or conventional, is composed of lobules of epithelioid cells separated by fibrous septa. These cells can be arranged in ribbons and cords in a myxoid extracellular matrix or as more dense collections of tumor cells. The tumor cells have vacuolated “bubbly” cytoplasm for which they are termed physaliphorous cells (▶**Fig. 28.2**a).[1]
     - Chondroid chordoma are tumors where the extracellular matrix resembles hyaline cartilage. Although there is some reported variability, these tumors are thought to behave in similar manner as chordoma NOS.[1]
     - Dedifferentiated chordoma refers to a tumor that has a biphasic pattern with features of chordoma NOS and undifferentiated spindle cell tumors or osteosarcomas.[1] This is the most aggressive pattern associated with early metastasis and very poor prognosis.[7]
       - On immunohistochemical analysis, chordomas are typically immunoreactive for cytokeratins,

## Answers (*continued*)

epithelial membrane antigen (EMA), S-100, and brachyury (▶ **Fig. 28.2b**).[1]

6. *What is the diagnostic marker of chordoma* (▶ **Fig.28.2b**)*?*
   - Brachyury, a nuclear transcription factor and product of the T-gene, is a marker for chordoma.[8]
7. *What are the common presenting signs and symptoms of clival chordomas?*
   - The presenting signs and symptoms vary among patients and depend on the location and size of the tumor as well as mass effect on adjacent neurovascular structures.
   - Patients with clival chordomas most commonly present with cranial neuropathies.[9,10] The most common presentation is diplopia from dysfunction of CN VI due to mid-clival involvement. For tumors confined to the upper clivus, CN III is most often involved, and for chordomas involving the lower clivus, lower CN palsies such as CN IX, X, and XII are common.[10] Other presenting signs and symptoms include headache, nasal obstruction/congestion, brainstem symptoms, endocrinopathies, hydrocephalus, and other cranial nerves deficits.
8. *What is the treatment of choice for patients with clival chordoma?*
   - Surgical resection followed by postoperative adjuvant proton beam radiation is the mainstay treatment for patients with clival chordoma.[11,12]
   - Postoperative proton beam radiotherapy is often used given the high dose of radiation required and the proximity to critical neural structures.
     - In a systematic review comparing various radiation modalities, the average 5-year local control and overall survival rates were 69.2 and 79.8%, respectively, after proton beam radiation; 36 and 53.5%, respectively, after conventional radiation therapy; 50 and 82%, respectively, after stereotactic fractionated radiation therapy; and 56 and 75%, respectively, after radiosurgery.[13]
9. *Discuss the various surgical approaches available to address this tumor.*
   - The surgical approach should take into consideration tumor location, extension, and relationship to adjacent neurovascular structures, including the internal carotid artery, cavernous sinus, cranial nerves, basilar artery, and brainstem. The goal of surgery is maximal tumor resection (optimally gross total resection) with preservation of neurovascular structures.
   - The endoscopic endonasal transclival approach is often used for patients with centrally occurring tumors.
   - Open approaches have been used traditionally to resect this tumor and several have been described, which are listed herein. For tumors that extend laterally, combined approaches are often required.[10,11]
     - The anterolateral approach or extreme lateral approach provides a good exposure from the mid clivus down to the upper cervical spine, and from one lateral side to beyond the midline contralaterally.
     - For more lateral extensions, the pterional transcavernous approach, the subtemporal transpetrous apex or infratemporal approach, the subtemporal transcavernous approach can be utilized.
     - For midline extension, the frontal transbasal approach, the extended transsphenoidal transmaxillary approach, the transfacial and transsphenoethmoidal approach, the midfacial degloving approach, the transoral approach, the mandible splitting approach, and the Le Fort I osteotomy approach have been used.[14,15]
10. *What anticipatory measures should be undertaken when planning open surgery on this tumor?*
    - Intraoperatively
      - One should take into consideration bony structures and stability of the craniocervical junction which must be respected or otherwise reconstructed via posterior fixation and fusion techniques (see spine section).
      - Cranial nerve function (especially lower CNs) should be preserved.
      - Bone drilling must be done meticulously with goals of preservation of the internal carotid artery and other adjacent neurovascular structures.
      - CSF leak is a common complication. One should be prepared to place a lumbar drain intraoperatively and plan for potential flap or graft placement for skull base reconstruction, should that become the case.
    - Perioperatively and postoperatively
      - Preoperative balloon test occlusion to assess vertebral and carotid artery involvement as well as collateralization is important to consider.
      - Swallowing difficulties and aspiration pneumonia can present a serious postoperative problem. A (temporary) tracheostomy should be discussed preoperatively with the patient.
      - Postoperative bone window CT scan and an MRI scan should be performed early to assess the extent of bone and tumor resection and stability of the craniocervical junction.
        General complications include infection, hematoma, trismus, malocclusion, cranial nerve deficits, spinal instability, injury to the brainstem or spinal cord, endocrine disturbances, stroke, internal carotid artery injury.[14,15]

## Answers (*continued*)

11. *What is the prognosis for patients with clival chordoma?*
    - There is some variability in the literature, however, in updated studies, the median survival for patients with clival chordoma treated with surgery and radiation is 9 years and the estimated 5- and 10-year overall survival rates are 70–80% and up to approximately 60%, respectively.[9,16,17]
    - Metastases to the lungs, liver, and bones can develop in some patients.[18]
12. *What is the role of chemotherapy in the management of patients with chordoma?*
    - Chordoma is a chemoresistant tumor.
    - There are no Food and Drug Administration (FDA) approved therapeutics for patients with chordoma.
    - There have been a few clinical trials evaluating systemic therapies.
      - A phase II study of 9-nitro-camptothecin, a topoisomerase 1 inhibitor, demonstrated an objective response in one out of 15 patients and a progression-free survival of 33% in patients with advanced chordoma.[19]
      - A phase II study of imatinib, a tyrosine kinase inhibitor active against platelet-derived growth factor receptors, c-KIT, and BCR-ABL, demonstrated median progression-free and overall survival rates of 9 and 35 months, respectively, in patients with advanced chordoma.[20]
      - A phase II study of lapatinib, a tyrosine kinase inhibitor active against epidermal growth factor receptor (EGFR) and HER2/neu, showed modest antitumor activity in patients with advanced chordoma.[21] Several case reports have also reported activity of other EGFR inhibitors.[22]
      - Clinical trials using a brachyury-based vaccine are underway.

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# Case 29 Petrous Apex Tumor

*Marc-Elie Nader, Shaan M. Raza, Franco DeMonte, Remi Nader, and Paul W. Gidley*

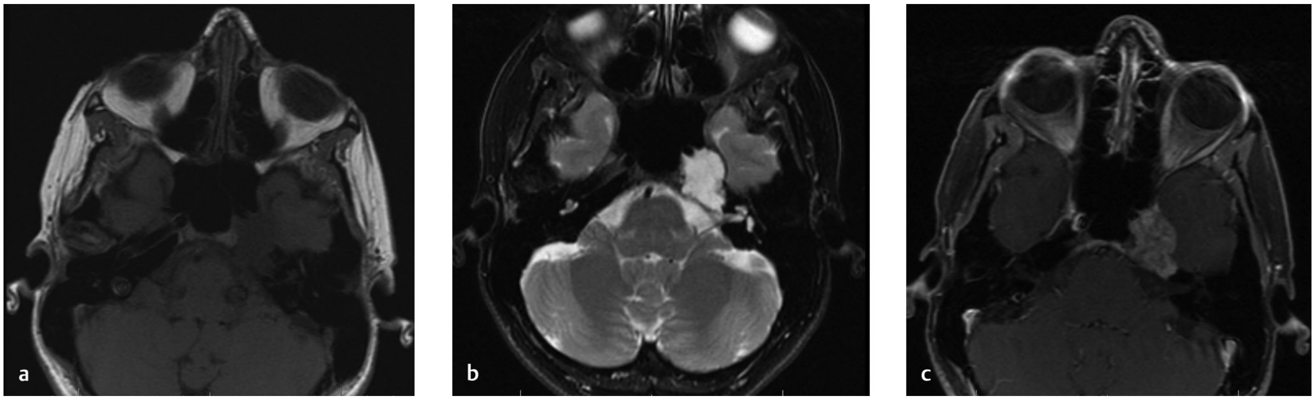

**Fig. 29.1** Preoperative axial MRI of the skull base showing a left petrous apex lesion. (**a**) T1-weighted sequence, (**b**) T2-weighted sequence, (**c**) T1-weighted sequence with contrast.

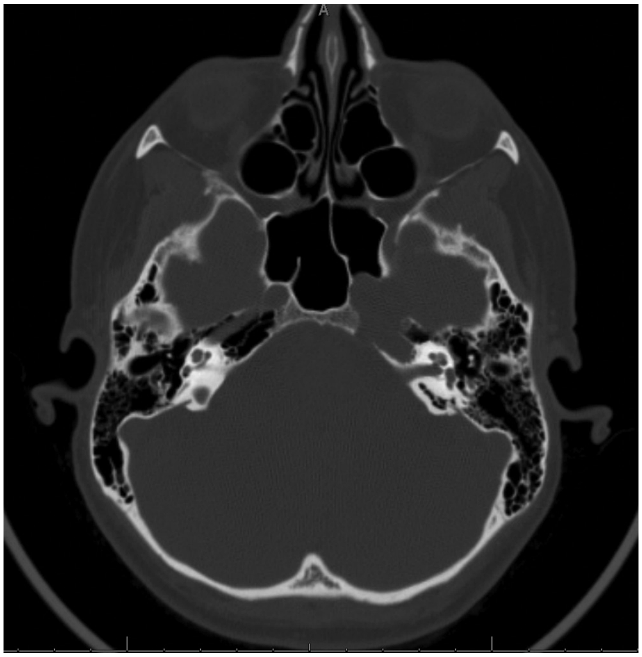

**Fig. 29.2** Preoperative axial CT scan of the skull base.

## Clinical Presentation

- A 42-year-old woman presents with 3-year history of diplopia on lateral gaze.
- Upon further questioning, the patient also reports episodes of left retro-orbital pain for the past 10 years.
- Neurologic examination reveals left-sided abducens nerve palsy and mild hypoesthesia in the distribution of the maxillary and mandibular divisions of the trigeminal nerve. No other focal deficits are seen.
- MRI and CT scans of the brain are shown in ▶**Fig. 29.1** and ▶**Fig. 29.2.**

## Questions

1. Interpret the CT scan.
2. Interpret the MRI.
3. Describe the course of the abducens nerve from the brainstem to the superior orbital fissure (SOF).
4. Give a differential diagnosis and the key radiologic findings for each diagnosis.
5. What is the most likely diagnosis and why?
6. What further studies should you obtain and which other medical services would you like to consult?
7. What are the management options in this case? The patient initially opted for observation with imaging at regular intervals. Her ocular symptoms initially improved with the use of prisms and strabismus surgery. Three years after initial diagnosis, the patient reported worsening of her diplopia. Repeat imaging showed tumor growth, and the patient opted for surgery.
8. Name the surgical approaches to the petrous apex.
9. Which factors influence the decision when choosing the surgical approach?
10. Discuss major complications associated with the extended middle fossa approach.

## Answers

1. *Interpret the CT scan.*
   - This axial view of the head in bone window shows a sharply circumscribed mass lesion involving the left petrous apex.
   - There is erosion of the medial margin of the carotid canal, the lateral aspect of the clivus, and the posterior aspect of the sphenoid sinus.
2. *Interpret the MRI.*
   - A lobular mass hypointense on T1-weighted images (▶**Fig. 29.1a**) and hyperintense on T2-weighted images (▶**Fig. 29.1b**) is seen involving the left petroclival region.
   - There is heterogeneous contrast enhancement (▶**Fig. 29.1c**). Anteriorly, the lesion abuts 180 degrees around the cavernous part of the left internal carotid artery (ICA).
3. *Describe the course of the abducens nerve from the brainstem to the superior orbital fissure (SOF).*
   - The abducens nucleus is located in the caudal pons ventral to the floor of the fourth ventricle.
   - The axons of the cranial nerve VI emerge at the medial aspect of the pontomedullary junction and travel laterally in an ascending trajectory toward the inferior petrosal sinus.
   - It then penetrates the meningeal layer of the sinus and travels within Dorello's canal in a medial to lateral and inferior to superior direction.
   - Eventually, it passes underneath Gruber's ligament and above the petrous apex to enter the posterior cavernous sinus.
   - In the cavernous sinus, it travels initially horizontal and lateral to the vertically oriented segment of paraclival ICA and then oblique, inferior to the parasellar segment of the ICA, and parallel to the ophthalmic segment of the trigeminal nerve until it reaches the SOF.
4. *Give a differential diagnosis and the key radiologic findings for each diagnosis.*
   - Noninfectious, nontumoral
     – Asymmetric pneumatization
     – Carotid aneurysm
     – Cholesterol granuloma
     – Cerebrospinal fluid (CSF) cyst/cephalocele
     – Epidermoid
     – Effusion
     – Mucocele
   - Infectious
     – Petrous apicitis
   - Benign tumors
     – Meningioma
     – Paraganglioma
     – Schwannoma
   - Malignant tumors
     – Chondrosarcoma
     – Chordoma
     – Hemangiopericytoma
     – Metastasis
     – Plasmacytoma/multiple myeloma
   - ▶**Table 29.1** summarizes the key radiologic findings for the most important diagnoses.
   - The main differentiator between chordomas and chondrosarcomas is the site of origin of the tumor. Chordomas originate centrally at the clivus while chondrosarcomas are located more laterally at the petroclival junction. Moreover, chondrosarcomas are more likely to exhibit matrix calcification.
5. *What is the most likely diagnosis and why?*
   - Chondrosarcoma, based on imaging:
     – The lesion is centered at the petrous apex
     – Bone erosion is present
     – It is hypointense on T1-weighted images, hyperintense on T2-weighted images, and enhances post contrast infusion
6. *What further studies should you obtain and which other medical services would you like to consult?*
   - Several steps are important for the work-up:
     – Imaging
       ○ CT without contrast to evaluate the degree of calcification of this lesion as well as the extent of erosion of the petrous apex and the clivus

**Table 29.1** Radiologic characteristics of petrous apex lesions

| | MRI | | | CT | Other |
|---|---|---|---|---|---|
| | T1 pre | T1 post | T2 | | |
| Cholesterol granuloma | + | − | + | Smooth erosion | |
| CSF cyst | − | − | + | | DWI − |
| Epidermoid | − | − | + | | DWI + |
| Mucocele | = | − | + | Destroyed septae | |
| Petrous apicitis | − | Rim enhancement | + | Destroyed septae | |
| Meningioma | =/+ | + | =/+ | Calcification | Dural tails, sessile |
| Paraganglioma | = | + | + | | Flow voids, salt/pepper |
| Schwannoma | = | + | −/+ | | Centered at IAC |
| Chordoma | −/= | + (less than chondrosarcoma) | + | Bone erosion | In clivus (central) |
| Chondrosarcoma | −/= | + | + | Bone erosion, matrix calcification | Centered at petrous apex |
| Metastasis | Variable | + | Variable | Bone erosion | Variable |

*Abbreviations:* CSF, cerebrospinal fluid; DWI, diffusion-weighted image; IAC, internal auditory canal; +, hyperintense; −, hypointense; =, isointense.

## Answers (*continued*)

  - ○ CT angiogram: to identify the direction of displacement of the ICA and its branches with respect to the lesion which will help in designing the surgical approach
- – Consultation requests
  - ○ Ophthalmology: a full preoperative evaluation is necessary
  - ○ Endocrinology: complete evaluation of the hypothalamic–pituitary axis is crucial as many of these lesions may compress the pituitary gland or its stalk. Furthermore, it is important to rule out nonsurgical lesions on the differential diagnosis such as giant prolactinomas.
  - ○ Otorhinolaryngology: in cases where the lesion is extending posteriorly toward the lateral part of the petrous apex and in cases where an endoscopic endonasal approach is entertained

7. *What are the management options in this case?*
   - The preferred treatment option is complete surgical resection of all invaded bone and any soft-tissue extension.[1]
   - Every effort is made to preserve vital structures, even if this entails subtotal removal.
   - The data regarding adjuvant radiation therapy is currently mixed. Current data indicates that adjuvant radiotherapy is administered based on histologic subtype and grade (all mesenchymal, all dedifferentiated, high-grade conventional tumors).
   - Observation may sometimes be considered. Reasons for initial observation include syndrome-associated tumor (i.e., possible enchondroma with Ollier's syndrome), small tumor size, minimal or no symptoms, older age, and advanced comorbidities.[2] The patient initially opted for observation with control imaging at regular intervals. Her ocular symptoms improved with the use of prisms and following strabismus surgery. Three years after initial diagnosis, the patient reported worsening of her diplopia. Repeat imaging showed tumor growth, and the patient opted for surgery.
8. *Name the surgical approaches to the petrous apex* (▶ **Fig. 29.3**).
   - Non-hearing preservation:
     - Translabyrinthine
     - Transcochlear/transpetrosal
   - Subtemporal approaches:
     - Standard middle fossa
     - Extended middle fossa
     - Kawase approach
   - Suboccipital/retrosigmoid
   - Combined approaches:
     - Retrolabyrinthine + middle fossa
   - Infratemporal
   - Endonasal endoscopic transpterygoid
9. *Which factors influence the decision of the surgical approach?*
   - Tumor location and relation to carotid artery
   - Intradural extension
   - Patient's comorbidities
   - Patient's neurologic deficits
   - Comfort of the surgeon with the approach
10. *The patient underwent surgery and the tumor was resected through an extended middle fossa approach. Discuss major complications associated with this approach.*
    - Brainstem injury
    - Cerebral edema: can be reduced by minimizing retraction time, using osmotic agents, and hyperventilation. Aggressive removal of bone of the anterior petrous apex also offers a wider surgical corridor, helping minimize the need to retract.

## Answers (*continued*)

- Cranial nerve deficits: trochlear, trigeminal, abducens and facial nerves
- CSF leak
- Hematoma
- Stroke
- Seizure: perioperative antiepileptics may be given and protection of any exposed brain can help reduce the risk of seizures[3,4]

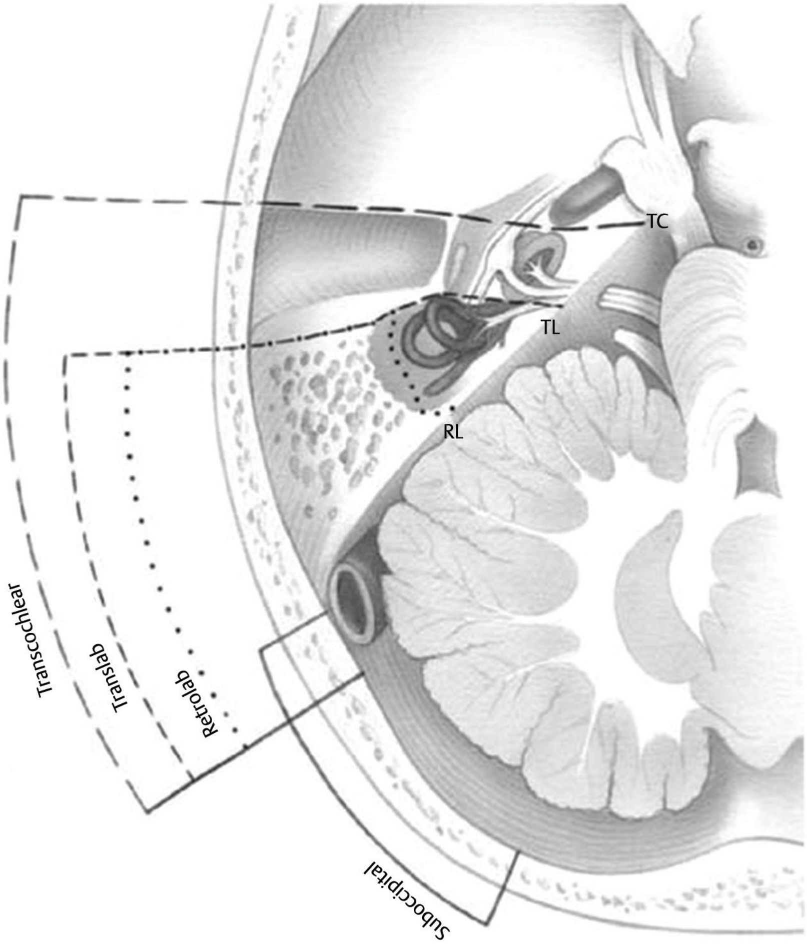

**Fig. 29.3** Axial view of the left petrous pyramid with four common approaches to the petrous apex. RL, retrolabyrinthine; TC, transcochlear; TL, translabyrinthine. (Reproduced from Jackler RK. Atlas of Skull Base Surgery and Neurotology. New York: Thieme Medical Publishers: 2009.)

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# Case 30 Intracranial Chondrosarcoma

*Mohamed A. Labib, Hussam Abou-Al-Shaar, Abdul Haseeb Naeem, Mohammed Saeed Bafaqeeh, and Peter Nakaji*

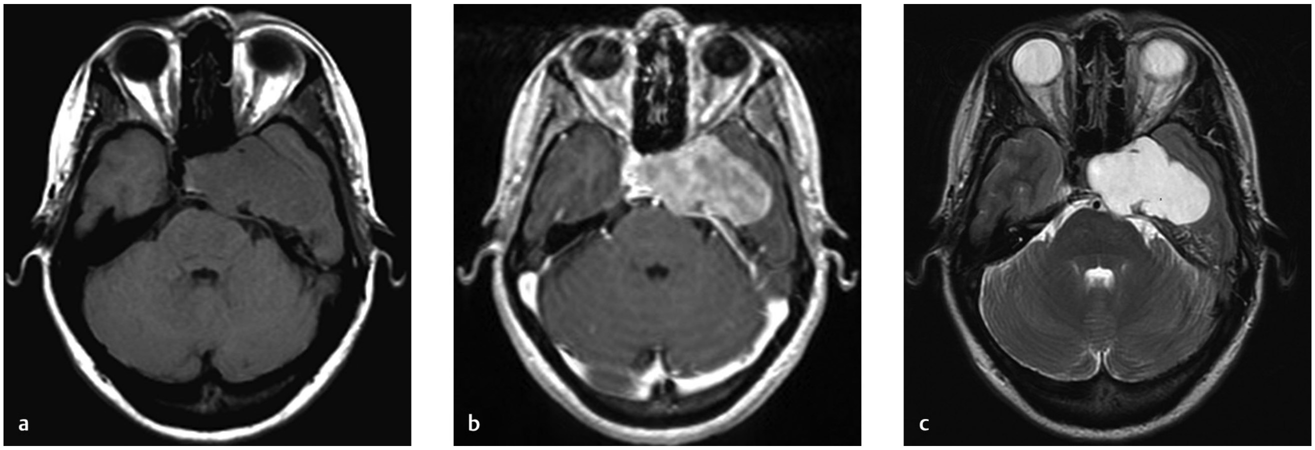

**Fig. 30.1** Axial MRI of the skull base showing a left petrous apex lesion. (**a**) T1-weighted sequence, (**b**) T1-weighted sequence with contrast, and (**c**) T2-weighted sequence.

## Clinical Presentation

- A 23-year-old male university student was referred from the emergency department with a 3-week history of headaches and worsening diplopia.
- Initially, he attributed his symptoms to stress as he was going through his final exams.
- He reports worsening diplopia on left lateral gaze.
- His examination reveals left abducens palsy.
- A CT scan and MRI were performed (▶**Fig. 30.1**).

## Questions

1. Describe the important findings on the MRI scan.
2. List a few conditions that may be associated with chondrosarcoma.
3. What is the most common site for skull base chondrosarcoma?
4. List three surgical approaches for this lesion along with their advantages and disadvantages.
5. List the different subtypes of chondrosarcoma. Which one of the subtypes is most common in the cranial base?
6. List the different grades of chondrosarcoma. Which one is the most common?
7. List three theories for the development of chondrosarcoma.
8. What is the 5-year survival rate of patients with chondrosarcoma?
9. List three negative prognostic factors for chondrosarcoma.
10. What is the role of radiation therapy in the management of this tumor?
11. What is the role of chemotherapy in the treatment of chondrosarcomas?

## Answers

1. *Describe the important findings on the MRI scan.*
   - A large left petro-spheno-clival lesion is seen to have eroded part of the clivus. The lesion is high signal on T2-weighted images, isointense on T1-weighted images, and is heterogeneously contrast enhancing.
2. *List a few conditions that may be associated with chondrosarcoma.*
   - The majority of chondrosarcoma cases are sporadic; however, conditions that may predispose patients to developing these lesions include:
     - Ollier disease: enchondromas developing close to growth plate cartilage
     - Maffucci syndrome: enchondromas, hemangiomas, lymphangiomas
     - Paget's disease
     - Osteochondroma
3. *What is the most common site for skull base chondrosarcoma?*
   - The petroclival fissure: 66% of chondrosarcomas arise from the petroclival fissure, 28% from the clivus, and the remaining 6% arise from the spheno-ethmoidal complex.[1]
4. *List three surgical approaches for this lesion, along with their advantages and disadvantages.*
   - Anterior approaches, such as the endoscopic endonasal transclival, petrous apex, and far medial approaches,[2–4] allow direct access to the majority of lesions from the dorsum sella rostrally to the foramen magnum caudally. These approaches are particularly useful where the lower cranial nerves or the nerves of the cavernous sinus are displaced posteriorly or laterally.

     In a large study of 800 patients,[5] the morbidities associated with endoscopic endonasal approaches in general (not limited to chondrosarcoma patients only) included cerebrospinal fluid (CSF) leak (16%) and vascular injury (0.9%). With the adaptation of vascularized flaps for skull base reconstruction, the incidence of CSF leak dropped to less than 6%.[5]
   - Anterolateral approaches, such as the orbitozygomatic approach, provide access to parasellar area and the tip of the clivus. These also allow direct access to lesions involving the cavernous sinus.
   - Lateral approaches, such as the anterior petrosectomy approach, provide access to the anterior part of the posterior fossa and the upper third of the clivus. The middle third of the clivus can be accessed via a posterior petrosectomy (presigmoid retrolabyrinthine) approach with varying degrees of semicircular canal removal to expand the surgical corridor.
   - Finally, lesions occupying the lower third of the clivus can be addressed via a retrosigmoid or a more extensive far lateral approach in cases where a significant portion of the tumor is situated ventral to the brainstem.
5. *List the different subtypes of chondrosarcoma. Which one of the subtypes is most common in the cranial base?*
   - Conventional, mesenchymal, dedifferentiated, and clear cell are the four subtypes of chondrosarcoma.
     - Conventional chondrosarcomas account for almost all intracranial chondrosarcomas.
     - The mesenchymal subtype constitutes less than 10% of all skull base chondrosarcomas.
     - No reports to date of clear cell or dedifferentiated subtypes occurring intracranially, as these subtypes do not arise in the axial skeleton.[6,7]
6. *List the different grades of chondrosarcoma. Which one is the most common?*
   - Grading of chondrosarcoma follows the World Health Organization (WHO) histological grading system that takes into consideration the degree of cellularity, cytologic atypia, and mitotic activity of the tumor.
     - Grade I tumors are well-differentiated tumors.
     - Grade II represents a moderately differentiated histology.
     - Grade III demonstrates a poorly differentiated histological pattern.
     - Grade IV tumor is a completely undifferentiated tumor.
   - According to the largest series study of intracranial chondrosarcoma of 200 patients, 51% of patients had grade I tumors, 21% had grade II tumors, and 29% demonstrated a mixed grade I and II. Interestingly, no patients were reported to have grade III or IV tumors.[1,6]
7. *List three theories for the development of chondrosarcomas.*
   - The first theory relies on the presumption that bones of the skull base arise mainly by endochondral ossification.[8] It postulates that intracranial chondrosarcomas develop from the chondrocytes within rests of endochondral cartilage that reside around the region of foramen lacerum. This theory is particularly plausible given that chondrosarcoma is more common in the region of the petro-occipital, spheno-occipital, and sphenopetrosal synchondrosis.[9,10]
   - Another theory postulates that during the embryogenesis of the skull base and temporal bone, some of the primitive multifunctional pluripotent mesenchymal cells may contribute to the future development of chondrosarcomas.[10–12]
   - Finally, it has also been postulated that cranial base chondrosarcomas may arise from metaplastic changes of mature skull base fibroblasts.[10–12]

## Answers (*continued*)

8. *What is the 5-year survival rate of patients with chondrosarcoma?*
   - Progression-free survival/local control[13–16]
     - 5-year
       - Grade 1: 90%
       - Grade 2: 64%
       - Grade 3/mesenchymal/dedifferentiated: 34%
     - 10-year: 32 to 100%
   - Overall survival[13–16]
     - 5-year: 75 to 100%
     - 10-year: 71 to 100%
   - The overall 5-year mortality rate among all patients with skull base chondrosarcoma ranges from 0 to 15%.[13–17]
   - A recent systematic review on this topic showed that the 5-year mortality rate is closer to 11% with an average survival of time of 53.7 months.[10]
   - Long-term follow-up with MRI is necessary to detect late recurrence because tumor growth is generally indolent.[14]
9. *List three negative prognostic factors for chondrosarcoma.*

   Recent studies have identified the following negative prognostic factors:[1,10,18,19]
   - Mesenchymal subtype: the mortality rate in patients harboring the mesenchymal subtype was 54 versus 6% in patients with conventional chondrosarcoma.
   - High WHO grade: the mortality rates for grades I, II, and III were 5, 10, and 25%, respectively.
   - Lack of postoperative adjuvant radiation: the 5-year mortality rate was higher in patients who received surgery alone versus surgery and radiation therapy (25 vs. 9%).
10. *What is the role of radiation therapy in the management of this tumor?*
    - Radiation therapy is usually employed as an adjuvant treatment to surgery in patients with particular histologic subtypes, or in cases of significant involvement regardless of extent of resection.[20]
    - To date, there is no class I evidence suggesting the superiority of one radiation modality over the other. Below are two different options:
      - Particle (proton and carbon ion) radiotherapy:
        - Proton beam therapy (PBT) can deliver significantly high radiation doses to the target lesions while sparing “bystander” tissue. Because of the minimal exit dose after energy deposition to the target volume (Bragg peak) and the sharp lateral margins, critical structures, such as the brainstem and cranial nerves, can be spared. Furthermore, there is a theoretical radiobiological advantage of protons over photons. Clinically, progression-free survival and overall survival when adjuvant PBT is used following surgery ranges from 75 to 95% and from 85 to 95%, respectively.[7,21–24] PBT, however, is available in only a small number of centers worldwide.
        - Carbon ion particles have the similar energy characteristics to protons. Limited data, however, suggest a greater radiobiological effectiveness in the target tissue when compared to protons.[25] The limited availability of this treatment modality and its overall cost precludes its current use as a standard of care.
      - Stereotactic radiosurgery (SRS):
        - This modality has the advantage of delivering highly conformal large radiation doses to a tumor in a single setting (as opposed to fractionation as with proton therapy). Nonetheless, the surrounding tissue at risk may still receive higher doses of radiation than in case where protons are used. The largest multicenter study to date on the use of adjuvant SRS for the treatment of skull base chondrosarcoma has reported a 5-year progression-free survival and a 5-year overall survival of 86 and 85%, respectively.[20]
11. *What is the role of chemotherapy in the treatment of chondrosarcomas?*
    - The role of chemotherapy in the treatment of chondrosarcoma of the petrous apex is not well defined. Consideration can be given to neoadjuvant chemotherapy (i.e., chemotherapy protocols similar to that used in other high-grade sarcomas) for more aggressive (mesenchymal and dedifferentiated) subtypes and adjuvant chemotherapy for recurrent disease.[14]
    - Chemotherapy can be used in the palliative setting after multiple recurrences or in cases with distant metastases.

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# Section II Intracranial Pathology: Vascular Neurosurgery

# Case 31 Dural Arteriovenous Fistula

*Nancy McLaughlin and Michel W. Bojanowski*

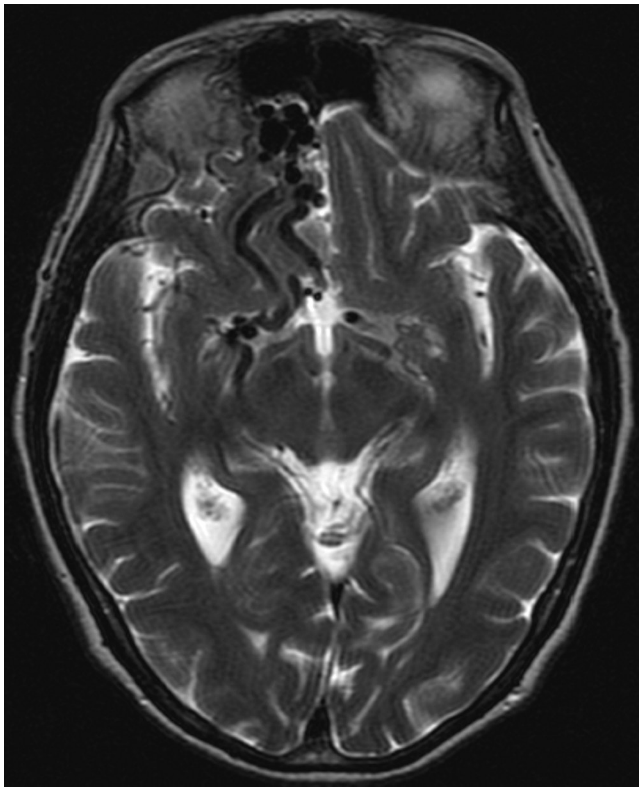

**Fig. 31.1** Axial T2-weighted MRI of the head.

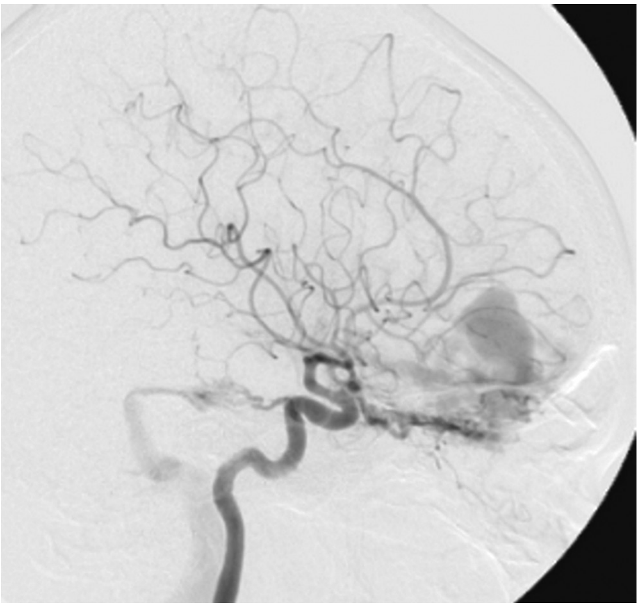

**Fig. 31.2** Digital subtraction angiography lateral view, right carotid injection.

## Clinical Presentation

- A 68-year-old woman was referred for evaluation of a pulsatile tinnitus ongoing for the past 7 years and a more recent headache of variable intensity occurring daily for 1 year. The patient is in otherwise excellent health. No other symptoms were reported.
- Neurological examination was normal; specifically, no bruit was auscultated over the skull and no pulsatile tinnitus was documented.

## Questions

1. What is your differential diagnosis for pulsatile tinnitus?
2. What are the indications to investigate a pulsatile tinnitus?
   Findings on the MRI scan (▶**Fig. 31.1**) revealed a vascular anomaly and required an angiography (▶**Fig. 31.2**).
3. Describe the angiography.
4. What is your management? Justify.
   You decided to treat the anterior cranial fossa dural arteriovenous fistula (DAVF) surgically.
5. What is the main step during surgery to eliminate the fistula?
6. What are the possible complications of surgical treatment?
   The postoperative angiography shows a complete exclusion of the anterior cranial fossa DAVF. However, there is now a retrograde venous drainage from the sigmoid sinus fistula (▶**Fig. 31.3**).
7. What is your management?
8. What are the causes of DAVFs?
9. How are DAVFs classified?

## Answers

1. *What is your differential diagnosis for pulsatile tinnitus?*
   - See ▶**Table 31.1** for details.[1,2]
2. *What are the indications to investigate a pulsatile tinnitus?*
   - In the absence of an audible bruit, MRI with MR angiography (MRA) is an appropriate initial diagnostic step for subjective pulsatile tinnitus.
   - In the presence of an objective pulsatile tinnitus, the clinician may proceed initially with an MRI-MRA. However, in patients with an audible bruit and those with a history of trauma accompanied with de novo pulsatile tinnitus, a conventional angiography is warranted, since it remains the most accurate method for detection of a DAVF.[1,3]
3. *Describe the angiography*.
   - The angiography showed a right anterior cranial fossa DAVF supplied by branches of the right and left ophthalmic arteries, right and left internal maxillary arteries, and right middle meningeal artery.
   - A large venous pouch located in the right anterior fossa was noted. Retrograde venous drainage to cortical veins toward the basal vein of Rosenthal and the great cerebral vein of Galen was present.
   - Other cortical veins refluxed toward the longitudinal superior sagittal sinus.
   - The angiography also revealed a right sigmoid sinus fistula nourished by the right tentorial artery. No retrograde venous reflux was noted (▶**Fig. 31.2**).
4. *What is your management? Justify*.
   - Treatment is indicated to eliminate the risk of hemorrhage and progressive neurological deficits. These risks are directly related to the presence of cortical venous retrograde drainage (CVRD). The overall morbidity and mortality rates of patients harboring a DAVF with CVRD are 15 and 10%, respectively.[4] A report has, however, suggested that asymptomatic DAVF with CVRD is associated with a less aggressive clinical course compared to symptomatic DAVF with CVRD (1.4 vs. 19% annual risk of neurological event).[5]
   - Anterior cranial fossa DAVFs always drain via cortical venous drainage and therefore, mandate an aggressive treatment.[6]
   - Endovascular embolization via transvenous or transarterial routes is the mainstay of DAVF treatment.[3, 7]
   - However, in this case, transarterial embolization is limited to branches of the external carotid artery. Embolization of ophthalmic arteries is avoided due to the inherent risk of central retinal artery occlusion.
   - Transvenous embolization is not feasible for most of these fistulas because of the lack of venous access.
   - Surgery is the best treatment option for anterior cranial fossa DAVFs.[8,9]
   - Lesions without CVRD, such as the sigmoid sinus fistula, are associated with benign natural history and do not require treatment except in case where symptoms are intolerable. In such situations, a palliative treatment may be indicated.[6] However, rarely such a fistula may develop a CVRD and any modification in patient's symptomatology should prompt further imaging investigation.[10]
5. *What is the main step during surgery to eliminate the fistula?*
   - The goal is to interrupt the draining veins at their dural origin.
6. *What are the possible complications of surgical treatment?*
   - Possible complications of surgical treatment of anterior fossa DAVFs include excessive blood loss, venous hypertension, venous infarct, cerebral edema, and seizures.
7. *What is your management?*
   - Prior to surgery of the anterior fossa DAVF, the sigmoid fistula was initially considered for conservative treatment, since there was no CVRD.
   - However, after surgery of the anterior fossa DAVF, the sigmoid fistula developed a retrograde venous drainage and a curative treatment for this fistula should be sought.
   - Transvenous embolization should be considered initially.
   - If endovascular treatment fails, surgery should be performed for patients with CVRD.
   - Whenever possible, transarterial embolization should be attempted to reduce intraoperative bleeding.[6]
8. *What are the causes of DAVFs?*
   - DAVFs are dural-based shunts supplied by pachymeningeal arteries, dural branches of leptomeningeal arteries, and/or the vertebral arteries, with a venous drainage via dural venous sinuses or via cortical veins.
   - The etiology of DAVFs remains poorly understood. Some conditions have been associated with DAVF such as head injury, prior craniotomy, infection, arterial dysplasia, and dural venous sinus thrombosis.
   - Most of these etiologies have a sinus occlusion in common.
9. *How are DAVFs classified?*
   - Digital subtraction angiography is the most accurate method for classification of DAVFs.[3]
   - The most commonly used classifications are those of Borden and colleagues[11] and Cognard and colleagues.[12] These classifications are based on the pattern of venous drainage of the arteriovenous fistulas (▶**Fig. 31.4** and ▶**Table 31.2**) and have been developed to predict their natural history.

**Table 31.1** Causes of pulsatile tinnitus

| Type of Lesion | Etiology |
|---|---|
| Arterial lesion | AVM |
| | Dural arteriovenous fistula |
| | Carotid cavernous fistula |
| | Aneurysm of the ICA |
| | Fibromuscular dysplasia of the ICA |
| | Dissection of ICA |
| | Aberrant carotid artery |
| | Atherosclerosis |
| | Vascular anomaly of the ear |
| | Vascular compression of the eighth nerve |
| Venous lesion | Jugular bulb anomalies (high-riding jugular bulb, dehiscent jugular vein, jugular diverticulum) |
| | Dominant or attenuated transverse sinus |
| | Benign intracranial hypertension |
| | Abnormal condylar or mastoid emissary veins |
| Neoplasm | Glomus jugular tumors |
| | Facial nerve hemangiomas |
| | Cavernous hemangioma |
| | Histiocytosis X |
| | Paget's disease |
| Miscellaneous | Anemia |
| | High cardiac outflow |
| | Thyrotoxicosis |
| | Otosclerosis |

*Abbreviations:* AVM, arteriovenous malformation; ICA, internal carotid artery.
*Source:* Adapted from Shin et al 2000; Weissman and Hirsch 2000.[1,2]

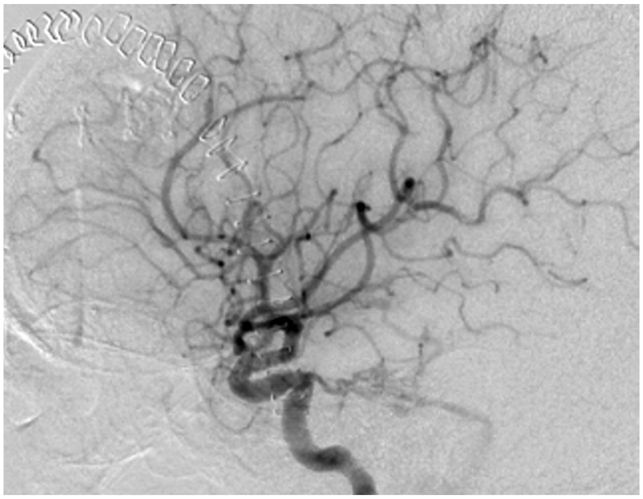

**Fig. 31.3** Digital subtraction angiography postoperative lateral view, right carotid injection.

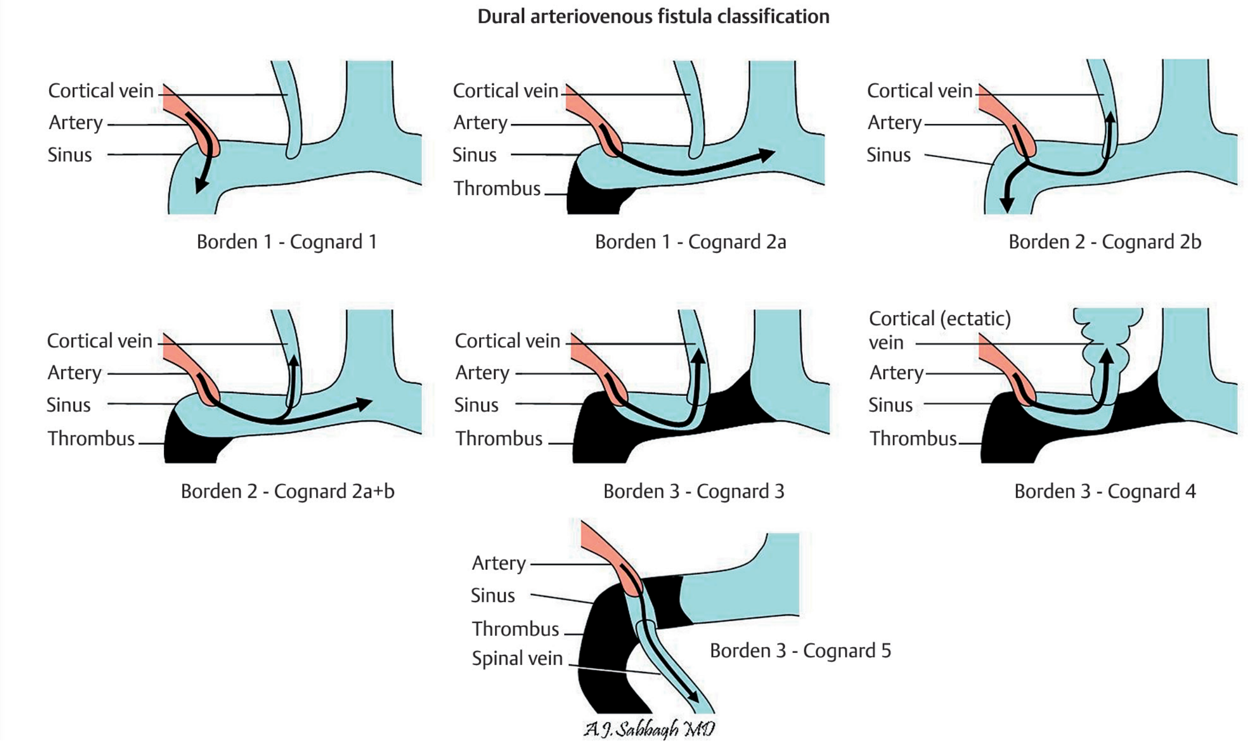
Dural arteriovenous fistula classification
Cortical vein
Artery
Sinus
Borden 1 - Cognard 1
Cortical vein
Artery
Sinus
Thrombus
Borden 1 - Cognard 2a
Cortical vein
Artery
Sinus
Borden 2 - Cognard 2b
Cortical vein
Artery
Sinus
Thrombus
Borden 2 - Cognard 2a+b
Cortical vein
Artery
Sinus
Thrombus
Borden 3 - Cognard 3
Cortical (ectatic) vein
Artery
Sinus
Thrombus
Borden 3 - Cognard 4
Artery
Sinus
Thrombus
Spinal vein
Borden 3 - Cognard 5
A.J. Sabbagh MD

**Fig. 31.4** Artist's rendering of venous drainage pattern of dural arteriovenous fistulas according to Borden and Cognard classification schemes. (Adapted from Borden et al 1995; Cognard et al 1995)[11,12]

**Table 31.2** Venous drainage pattern of dural arteriovenous fistulas according to Borden and Cognard classification schemes

| Borden Classification[11] | Cognard Classification[12] |
|---|---|
| Type I: drainage into dural venous sinus or meningeal vein only | Type I: drainage into dural venous sinus only, normal anterograde flow |
| Type II: drainage into dural venous sinus or meningeal vein + cortical venous reflux | Type IIa: drainage into dural venous sinus only, with retrograde flow |
| Type III: cortical venous reflux only | Type IIb: drainage into dural venous sinus (anterograde flow) + CVR |
| | Type III: CVR only without venous ectasia |
| | Type IV: CVR only with venous ectasia |
| | Type V: drainage into spinal perimedullary veins |

*Abbreviation:* CVR, cortical venous reflux.
Source: Adapted from Borden et al 1995; Cognard et al 1995.[11,12]
Note: All three types of dural arteriovenous fistulous malformation (AVFM) are subclassified as subtype a: simple fistula, and subtype b: multiple fistulas with multiple dural-based AVFMs fed by multiple arteries.

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# Case 32 Cerebral Arteriovenous Malformation

*Badih Daou, Pascal M. Jabbour, and Erol Veznedaroglu*

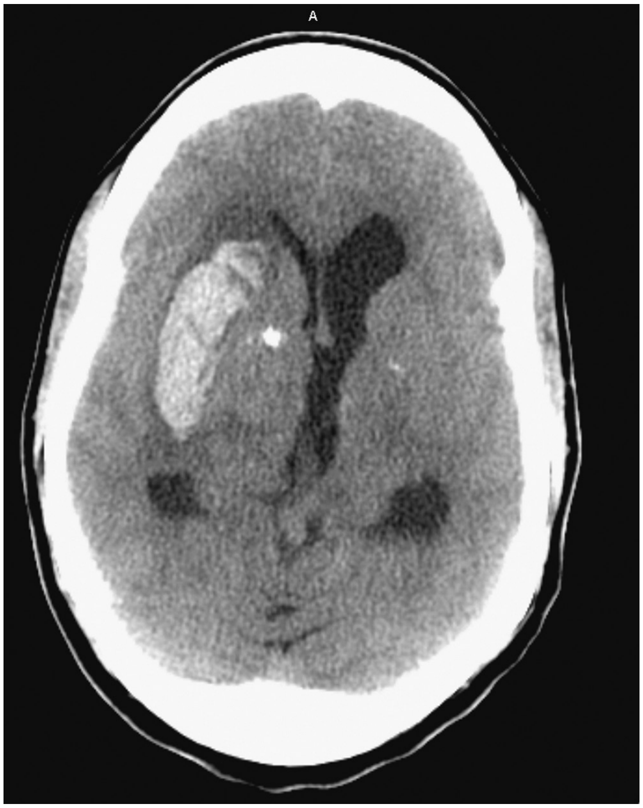

**Fig. 32.1** CT scan of the head showing a right basal ganglia bleed.

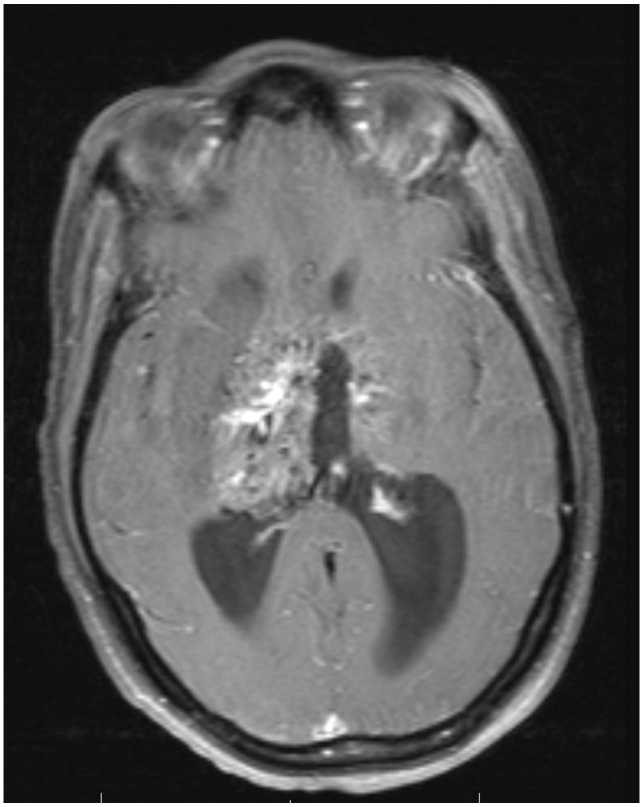

**Fig. 32.2** MRI scan of the head showing a basal ganglia arteriovenous malformation.

## Clinical Presentation

- A 36-year-old woman presents to the emergency room for sudden onset of headaches and right hemiplegia.
- Her medical and familial history was noncontributory.
- Neurologic evaluation showed that the patient was awake, alert, and strength was 5/5 on the right; she was hemiplegic on the left, with right-sided gaze preference, anosognosia, and hemiasomatognosia.
- CT scan is shown in ▶**Fig. 32.1** and MRI scan of the head is shown in ▶**Fig. 32.2**.

## Questions

1. Considering the age of the patient, what is the most likely etiology of the bleed?
2. What is the next test to order?
3. What is the most common arteriovenous malformation (AVM) grading system used and what is the patient's grade? What are some other classification systems?
4. What is the risk of rupture of an AVM and the lifetime cumulative risk for this patient?
5. What are the different treatment options for this patient?
6. What are some general treatment approaches?
7. What are some important steps of AVM surgery?
8. What are the risks associated with AVM treatment?
9. What is normal perfusion pressure breakthrough and how is it treated?

## Answers

1. *Considering the age of the patient, what is the most likely etiology of the bleed?*
   - Any intracerebral hemorrhage in a young patient without any past medical history should raise the suspicion of a vascular malformation.
2. *What is the next test to order?*
   - The CT scan of the head showed a basal ganglia bleed in a young patient; the next test to be ordered should be an imaging modality that is able to demonstrate any vascular abnormality, such as a CT angiogram, MRI, or MR angiography.
3. *What is the most common AVM grading system used and what is the patient's grade? What are some other classification systems?*
   - The Spetzler-Martin grading system (▸**Table 32.1**)[1]
   - The patient's grade is 2 (size) + 1 (eloquence) + 1 (deep venous drainage) = 4
   - One modification of the Spetzler-Martin grading system divides grade III AVMs into four different combinations: small-deep-eloquent (S1V1E1), medium-deep (S2V1E0), medium-eloquent (S2V0E1), and large (S3V0E0).[2]
   - Another modification includes a system that consists of three classes of AVM by combining Spetzler-Martin grades I and II AVMs into class A and grades IV and V lesions into class C; grade III AVMs become class B.[3]
   - The supplementary Spetzler-Martin grading scale includes three other variables: age, hemorrhagic presentation, and nidal diffuseness (a compact AVM is wound tightly with distinct margins whereas a diffuse AVM is wound loosely, with indistinct margins and poor separability).[4] When used along with the Spetzler-Martin grading system, the supplementary grading system has been shown to have higher predictive accuracy with improved preoperative risk prediction.
4. *What is the risk of rupture of an AVM and the lifetime cumulative risk for this patient?*
   - The risk of rupture of this AVM is ~4% per year.[5]
   - Risk is [1 − (risk of not hemorrhaging)] raised to the power of years left to live.
   - In a 36-year-old patient, at an average life expectancy of 79 years, the years left to live can be calculated to be 43 years.
   - Risk = $1 - (1 - \text{risk of hemorrhage})^{43}$
   - Risk = $1 - (0.96)^{43} = 83$
   - The patient has a lifetime risk of hemorrhage of 83%.
5. *What are the different treatment options for this patient?*
   - The different treatment options are:[1,6,7]
     - No intervention if the risk of intervening is higher than the natural history risk of the AVM rupturing
     - Embolization in preparation for surgical resection. Ideal targets for endovascular embolization are large grade 3 or 4 AVMs that can be embolized down to microsurgically accessible targets. Liquid embolic agents are the most commonly used materials to manage AVMs, including N-butyl cyanoacrylate (NBCA; Trufill, Codman, Raynham, Massachusetts, USA) and Onyx liquid embolic agent (Covidien, eV3 Neurovascular, Irvine, California, USA) which is an ethylene vinyl alcohol polymer dissolved in dimethyl sulfoxide (DMSO).[4] Platinum embolic coils can be concomitantly used.
     - Embolization with the goal of reducing the volume of the AVM in preparation for radiosurgery or embolization of AVMs that persist after radiosurgery
     - Surgery alone
     - Radiosurgery alone with possible volume fractionation. AVMs smaller than 3 cm are candidates for obliteration with radiosurgery.
6. *What are some general treatment approaches?*
   - Grade I and II AVMs are the best surgical targets if they are surgically accessible with low risk.[1] Radiation therapy (RT) alone can be used for grade I or II lesions if the AVM is < 3 cm in size and surgery has an increased risk based on location and vascular anatomy, considering that the patient will still be exposed to the risks of the AVM for 2 to 3 years following RT as the lesion gradually obliterates. Curative embolization of small AVMs with a low number of arterial feeders is an option.
   - Grade IV and V AVMs are often not amenable to surgical treatment alone because of the high procedural risk. A multimodal approach with a combination of embolization, radiosurgery, and/or surgery can be used.
   - Grade III AVMs remain potential targets for microsurgical resection but may require adjunctive embolization. Grade III lesions are generally treated with microsurgery or stereotactic radiosurgery, often with adjunctive endovascular therapy.
   - Small grade III AVMs are low-risk lesions and treated surgically. Medium/deep grade III lesions are of intermediate risk and careful selection for microsurgery is indicated. Medium/eloquent grade III AVMs are managed as high-risk lesions. Large grade III AVMs are rare and their surgical risk remains unclear, but surgery with endovascular treatment may be a possible treatment modality.
7. *What are some important steps of AVM surgery?*
   - Furosemide + mannitol is administered prior to dural opening.
   - Proximal temporary occlusion can be performed with aneurysm clips.

## Answers (*continued*)

- Complete sequential elimination of the arterial supply is performed first, and then the nidus is resected. The draining veins are ligated last to avoid increasing pressure in the nidus which can lead to intraoperative hemorrhage.
- Large draining veins are clipped with large aneurysm clips.
- Do not commit on vessel occlusion until the vessel is seen to enter the AVM.
- "If it looks like it may be AVM, it probably is."
- Difficult-to-control bleeding during this dissection is commonly an indication that the AVM–brain interface has been breached on the side of the AVM.
- As a safety measure, before taking the major draining vein, an aneurysm clip is placed across it for 10–15 minutes to test out the potential changes in the hemodynamics of the nidus and increase safety of the removal.
- Always obtain a postoperative angiogram and CT scan to evaluate for residual shunting.[8]

8. *What are the risks associated with AVM treatment?*
   - Endovascular embolization
     - Ischemic and hemorrhagic stroke resulting in transient or permanent neurological deficits.
     - This can be the result of arterial dissection or perforation by the guidewire or microcatheter, transition of a large amount of the embolic agent on the venous side of the AVM and immediate rupture, rupture of an associated aneurysm, vascular injuries during retrieval of the catheter and embolic material resulting in inadvertent occlusion of normal cerebral vasculature.
   - Microsurgical resection
     - Edema from retraction
     - Intraoperative rupture
     - Resection of normal brain tissue
     - Feeding vessel thrombosis
   - Radiosurgery
     - Risk of bleeding during the latency of 2 to 3 years
     - Neurologic deficits from edema and necrosis of normal brain tissue
     - Individual sensitivity to radiation
     - Delayed cyst formation
9. *What is normal perfusion pressure breakthrough and how is it treated?*
   - Normal perfusion pressure breakthrough is a disorder of autoregulation of the brain vasculature that is potentially seen after successful removal of an AVM and may present with sudden onset of brain swelling and bleeding from multiple sites.
   - Treatment
     - Elevation of the patient's head
     - Administration of mannitol and furosemide
     - Barbiturate- or etomidate-induced coma
     - Hypotension may be beneficial in the early postoperative period, up to a week, in patients who undergo surgery for the removal of an AVM of grade III or higher.

**Table 32.1** The Spetzler-Martin grading system[1] and supplementary grading scale[4]

| Spetzler-Martin Grading System | | | | | |
|---|---|---|---|---|---|
| **Size of AVM** | | **Eloquence of Adjacent Brain** | | **Pattern of Venous Drainage** | |
| Small (< 3 cm) | 1 | Noneloquent | 0 | Superficial only | 0 |
| Medium (3–6 cm) | 2 | Eloquent | 1 | Deep component | 1 |
| Large (> 6 cm) | 3 | – | | – | |
| **Supplementary Grading System** | | | | | |
| **Age** | | **Bleeding** | | **Compactness** | |
| < 20 | 1 | Yes | 0 | Yes | 0 |
| 20–40 | 2 | No | 1 | No | 1 |
| > 40 | 3 | – | | – | |

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# Case 33 Supratentorial Cavernous Angioma

*Julius July and Eka Julianta Wahjoepramono*

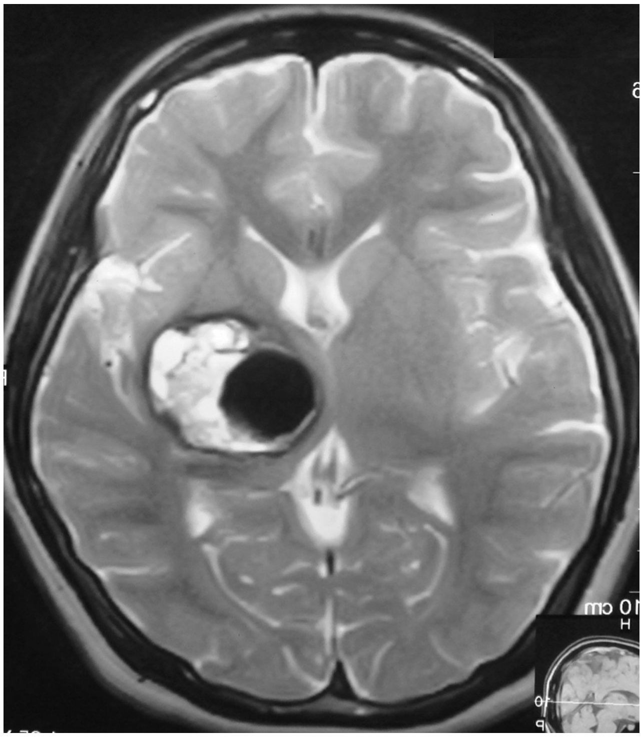

**Fig. 33.1** T2-weighted MRI showing a lesion involving the right basal ganglia and thalamus.

## Clinical Presentation

- A 19-year-old woman presents with chronic headache since early childhood. For the past 6 months her headaches have become progressively worse. She also felt that her left side was becoming weaker.
- There is no history of seizure.
- On initial assessment, she has left-side weakness and left hemihypoesthesia.
- She was referred from another hospital with the MRI study shown (▶**Fig. 33.1**).

## Questions

1. Describe the MRI feature of this lesion.
2. Provide a general classification scheme for intracranial vascular malformation.
3. What are the treatment options?
4. What treatment would you recommend for this case?
5. What is your argument between a surgical and nonsurgical option?
6. If you decide to do the surgery, what is your approach to remove the lesion?
7. What are the potential complications of surgery?

## Answers

1. *Describe the MRI feature of this lesion.*
   - Considering the clinical presentation and based on the T2-weighted MRI, the lesion demonstrates some acute or chronic hemorrhage.
   - The reticulated core of decreased and increased intensity with prominent surrounding rim of reduced intensity likely represent hemosiderin from previous hemorrhage.
   - The most likely diagnosis is of a cavernoma. Although unlikely, spontaneous hemorrhage and intratumoral bleeding should be on the differential diagnosis.[1,2]
2. *Provide a general classification scheme for intracranial vascular malformation.*
   - Intracranial vascular malformations are divided into following types:
     - Arteriovenous malformation (AVM)
     - Venous malformation
     - Capillary malformation
     - Cavernous malformation (cavernoma = cavernous angioma = cavernous hemangioma)[1,2]
3. *What are the treatment options?*
   - Conservative treatment is considered in the following cases:
     - Asymptomatic or minimally symptomatic lesions. This is based on the fact that these lesions may remain stable for a very long period and even if bleeding occurs, this tends to be mild given the low-pressure nature of the lesion.[3]
     - Multiple supratentorial cavernomas may be sporadic or familial. Sporadic cavernomas have been associated with history of radiation therapy to the brain and venous developmental anomalies.[4,5] The familial form can be as prevalent as 6 to 50% in some series.[6–8] These have an autosomal dominant inheritance of a mutation at one of the three cerebral cavernous malformation (CCM) loci, at chromosome 7q (*CCM1*), 7p (*CCM2*), and 3q (*CCM3*).[9,10]
     - Deep or eloquent areas where the surgical risks exceed the benefit, especially in elderly patients
   - Radiosurgery
     - In cases of surgically inaccessible lesions with at least two prior hemorrhages
     - Several studies show that radiosurgery could reduce the risk of hemorrhage to one-third of initial risk and also reduce the risk of seizures associated with it.
     - However, the benefits of radiosurgery are difficult to assess because of the unclear natural history of cavernomas, the inability to evaluate the status of the malformation vessels, and the lack of completeness of obliteration of the malformation.
   - Surgery
     - Recurrent hemorrhage, especially at a young age
     - Progressive neurologic deficits
     - Intractable epilepsy when benefits outweigh risks
     - Lesion located in the cerebellum or the cerebral cortex
     - Lesions that do not respond to radiosurgery
     - In special circumstances (e.g., a young woman who wants to become pregnant with an accessible lesion)
     - To prevent future bleeding[2,11]
4. *What treatment would you recommend for this case?*
   - Considering the patient's young age and history of repeated hemorrhage with progressive neurologic deterioration, surgical treatment should be recommended.
5. *What is your argument between a surgical and non-surgical option?*
   - Kondziolka et al[12] published a prospective study of 122 cavernomas that showed that the annual bleeding rate for symptomatic lesions is 4.5%.
   - The patient has a 93.6% chance of sustaining another hemorrhage in her lifetime (with life expectancy of 79 years, based on formula from Case 32, Cerebral Arteriovenous Malformation. The hemorrhage could be catastrophic.[1]
   - Although the lesion is located in eloquent brain (thalamus-basal ganglia), surgical resection still provides significant benefits that are greater than the risk involved.
6. *If you decide to do the surgery, what is your approach to remove the lesion?*
   - Based on the MRI (▶**Fig. 33.1**), the sylvian fissure appears to be quite relaxed. Resection can be achieved with image-guided surgery by performing a sylvian fissure dissection. A corticotomy along the insular gyrus of ~1 to 2 cm is enough to remove the cavernoma (▶**Fig. 33.2** and ▶**Fig. 33.3**).
   - Alternatively, a parietal transcortical-transventricular approach may also be used.
7. *What are the potential complications of surgery?*
   - The complications of surgery may include general risk that might happen during any neurosurgical cases (e.g., infection, cerebrospinal fluid leak, seizure, stroke, deep vein thrombosis, pneumonia, coma, death, etc.).
   - They also include neurologic worsening related to the location of the lesion and include paraesthesias, hemiparesis, or hemiplegia and visual field deficits, etc. [11,12]

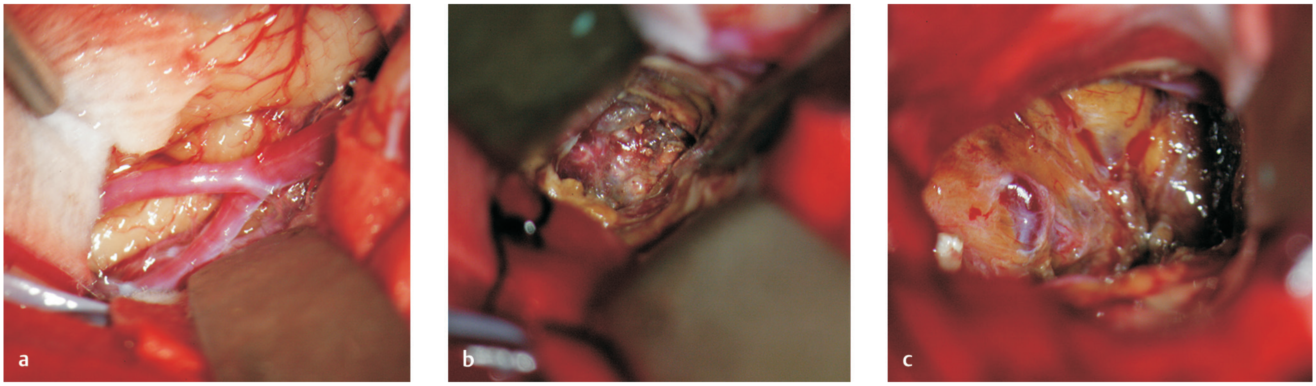

**Fig. 33.2** (**a**) Intraoperative photograph (300×) showing a sylvian fissure dissection, exposing the M2 segment of the right middle cerebral artery. Further dissection of the M2 segment provides maximal exposure of the long insular gyrus. (**b**) Intraoperative photograph (300×) taken after corticotomy completed at the long insular gyrus. The typical appearance of a cavernoma is situ is visualized. Very often, there is a good cleavage plane between the lesion and the surrounding brain. The surrounding brain has also undergone gliotic changes which facilitates complete resection. (**c**) Intraoperative photograph taken after complete removal of the lesion.

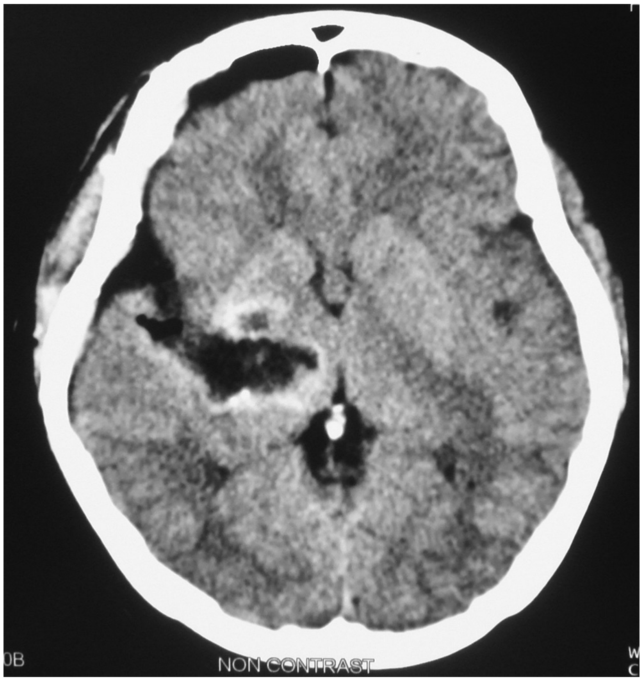

**Fig. 33.3** Postoperative CT scan showing the surgical track through the sylvian fissure.

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# Case 34 Brainstem Vascular Lesions

*Filippo Gagliardi, Alfio Spina, Michele Bailo, Cristian Gragnaniello, Alberto L. Gallotti, Anthony J. Caputy, and Pietro Mortini*

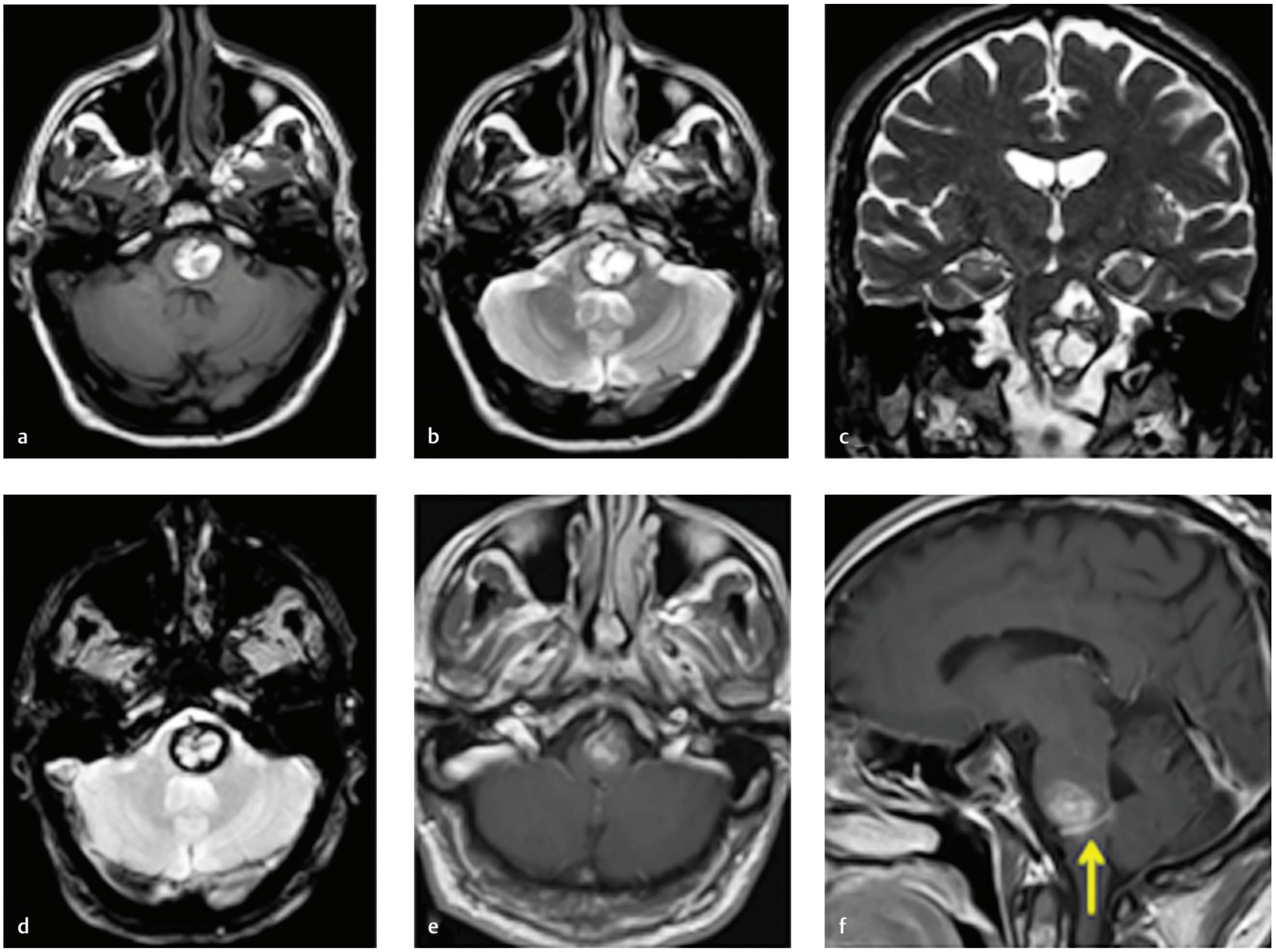

**Fig. 34.1** Preoperative MRI. (**a**) T1-weighted images, axial plane; (**b**) T2-weighted images, axial plane; (**c**) T2-weighted images, coronal plane; (**d**) gradient echo T2-weighted images, axial plane showing surrounding rim of hemosiderin; (**e**) T1-weighted images after contrast administration, axial plane; (**f**) T1-weighted images after contrast administration, sagital plane showing the presence of a developmental venous abnormality (*arrow*).

## Clinical Presentation

- A 55-year-old male suddenly experienced dysesthesia and weakness in the right arm, with subsequent onset of right hemiparesis and diplopia one week thereafter.
- The brain CT scan study showed a pontine hematoma.
- One month following a partial and transitory recovery, the patient was referred to our institution due to further clinical worsening.
- The neurological examination, performed at the time of admission, revealed a left 6th cranial nerve palsy, right 7th cranial nerve deficit (grade II according to House and Brackmann score) and a moderate right hemiparesis.
- The patient underwent CT scan showing a new brainstem hemorrhage.
- A cerebral MRI with DTI sequences and digital subtraction angiography (DSA) were performed to better define the lesion (▶**Fig. 34.1**).
- Patient underwent surgical resection through a retrosigmoid approach. Radical resection of the lesion was achieved, with a favorable postoperative neurological outcome (▶**Fig. 34.2**).

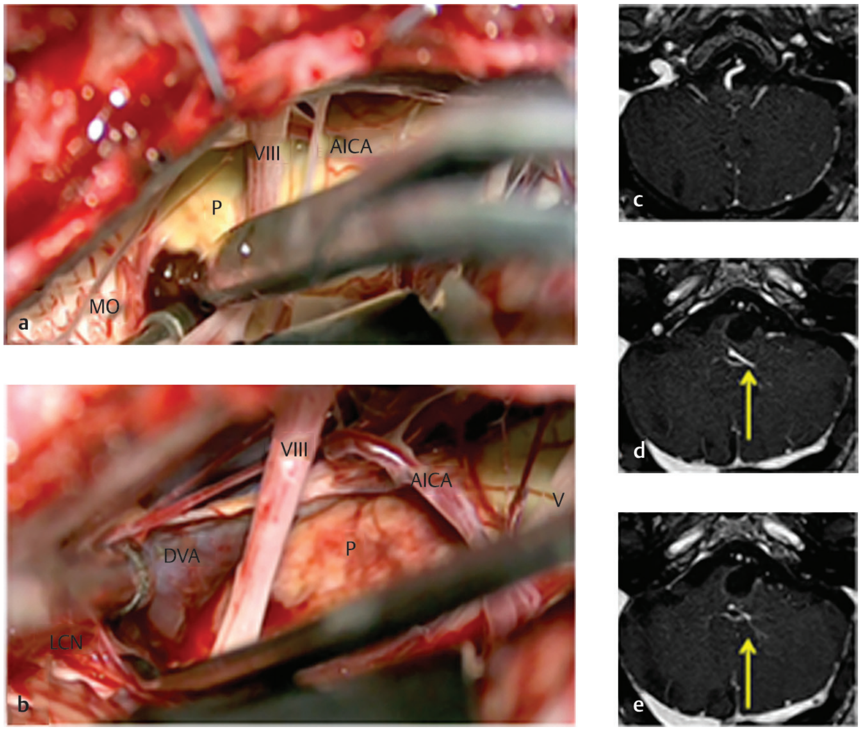

**Fig. 34.2** Intraoperative images and postoperative neuroradiological control. (**a**) Resection of the pontine cavernous hemangioma (CH); (**b**) Operative field after radical removal with preservation of the associated DVA; (**c–e**) postoperative MRI control, T1-weighted axial planes after contrast administration, showing DVA preservation (*arrows*) and confirming CH radical removal. AICA, anterior inferior cerebellar artery; DVA, developmental venous abnormality; LCN, lower cranial nerves; MO, medulla oblongata; P, pons; V, fifth cranial nerve; VIII, eight cranial nerve.

## Questions

1. Which are the anatomical bases of patient's clinical symptoms?
2. How would you manage this patient?
3. Describe the MRI study.
4. Explain the role played by gradient echo MRI images and DTI sequences in the diagnostic approach.
5. Is DSA useful in detecting cavernous hemangiomas (CH)?
6. How do CHs occur?
7. Why are the possible CH- associated developmental venous abnormalities (DVA) important in planning surgical resection of brainstem CH?
8. Which are the surgical approaches available to access this anatomical region?
9. Which are the treatment options for brainstem CH?
10. Which is the role played by radiosurgery in the treatment of CH?

## Answers

1. *Which are the anatomical bases of patient's clinical symptoms?*
   The patient experienced a hemorrhage within the anterior and rostral portion of the pontomedullary junction, where the following critical structures are located (▶**Fig. 34.3**):
   - The nucleus of the 6th cranial nerve: It is located in the caudal portion of the pons and projects into the floor of the fourth ventricle.
   - 7th cranial nerve fibers: The nucleus of the 6th cranial nerve is covered by bridging fibers of the 7th cranial nerve, departing from the facial nucleus and making a bulge along the ventricular floor, making the facial colliculi.[1]
   - The corticospinal tract: It runs through the anterior portion of the pons.[1]
2. *How would you manage this patient?*
   The first thing to do to evaluate the patient is to perform a CT scan and subsequently an MRI because of the following reasons:
   - CT scan: It may detect hemorrhage and define the location of the bleeding.
   - MRI: It confirms and better defines the type of hemorrhage, its anatomical location, morphologically characterizing the lesion, and its relationship with brainstem critical structures. Furthermore, MRI may show the possible presence of DVA, often associated with CH.

## Answers (*continued*)

- DSA: It may exclude other causes of bleeding (e.g., arteriovenous malformations and fistulas, as well as aneurysms).
- Neurophysiologic work-up: To preoperatively assess residual function of the corticospinal tracts and cranial nerves, a complete neurophysiologic work-up should comprise motor and somatosensory evoked potentials as well as auditory evoked potentials.

3. *Describe the MRI study.*
   - The MRI shows an intra-axial hemorrhagic lesion within the rostral anterior portion of the pontomedullary junction, visible as primarily hyperintense signal on both T1- and T2-weighted images.
   - The hemorrhagic lesion is well circumscribed, associated with surrounding hemosiderin deposits. It is circular in shape.
   - There also seems to be regions of “older” blood and “newer” blood within the lesion, giving it a characteristic “salt and pepper” appearance.
   - After contrast administration, an associated DVA surrounding the inferior right surface of the lesion is visible (▶**Fig. 34.1**).
4. *Explain the role played by gradient echo MRI images and DTI-sequences in the diagnostic approach.*
   - Gradient echo T2-weighted acquisitions are more sensitive in detecting the hemosiderin ring of CH.[2,3]
   - Diffusion tensor imaging (DTI) study is useful in mapping fiber projections to the sensorimotor cortical areas (▶**Fig. 34.1**).
5. *Is DSA useful in detecting CH?*
   - DSA is useful to rule out other vascular malformations such as aneurysms or arteriovenous malformations.
   - CH is considered to be an angiographically occult lesion: it is usually not visualized on DSA.[3]
6. *How do CHs occur?*
   - CHs can be detected as sporadic or, more rarely, familial cases. A familial case is based on an autosomal dominant trait.[2,3]
   - Solitary CHs occur three times more often than multiple ones. Familial cases typically present with multiple lesions, whereas nonfamilial cases usually present with only a single CH.
   - Three foci have been identified in the inherited form of CH, namely *CCM1* (cerebral cavernous malformation [CCM]) on chromosome 7q, *CCM2* on 7p, and *CCM3* on 3q. *CCM1* accounts for about half of inherited cases.
   - Nonfamilial cases of multiple CH are believed to occur in the following settings:
     - Unrecognized familial CH types
     - Multiple CHs associated with a single DVA
     - Multiple CHs following craniospinal irradiation
7. *Why are possible CH- associated DVA important in planning surgical resection of brainstem CH?*
   - DVAs are venous structures dedicated to the normal draining system; for this reason, they have to be preserved.
   - The cauterization and division of these structures can lead to brainstem venous infarction.[4–6]
8. *Which are the surgical approaches available to access this anatomical region* (▶**Fig. 34.2**)*?*
   - Suboccipital, retrosigmoid approach[7,8]
   - Anterior transpetrosal approach (Kawase)[8,9]
   - Presigmoid-retrolabyrinthine approach[8,10]
   - Midline posterior fossa approaches
9. *Which are treatment options for brainstem CH?*
   CH treatment options depend on lesion characteristics and clinical features.[3,4,6,11–13] A summary of treatment options as per the authors’ opinion is given below.
   - Asymptomatic, radiologically stable or nonhemorrhagic lesions:
     - Observation and neuroradiological follow-up by MRI
   - Epileptic lesions:
     - Observation and neuroradiological MRI follow-up
     - Antiepileptic medications for symptomatic control of seizures
   - Symptomatic, hemorrhagic, growing, or epileptic drug-resistant accessible lesions or patient's refusal:
     - Surgical resection
   - Deep-seated lesions or patients not eligible for surgery because of general medical conditions:
     - Radiosurgery
10. *Which is the role played by radiosurgery in the treatment of CH?*
    - Radiosurgery plays a role in decreasing the rate of rebleeding in the lesions that are considered surgically inaccessible or in patients who are not eligible for surgery.[11,13]
    - However, this modality is controversial as it does not completely eliminate the risk of rehemorrhage and may possibly cause radiation-induced complications.

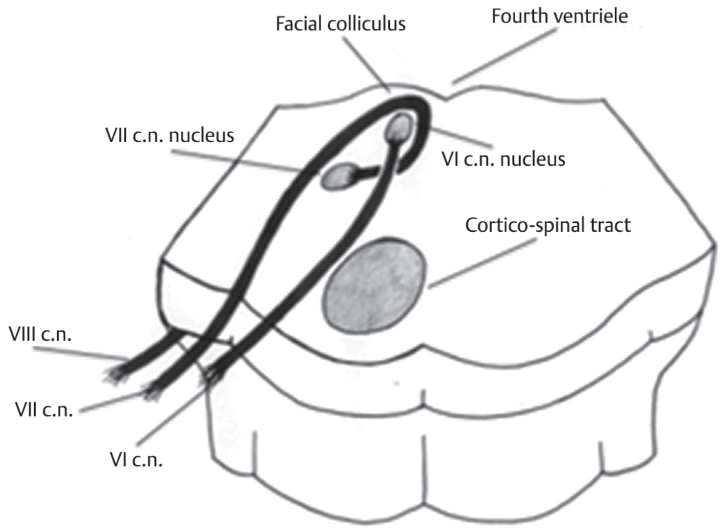

**Fig. 34.3** Schematic drawing depicting local anatomy. c. n., cranial nerve.

## Suggested Readings

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# Case 35 Carotid Cavernous Sinus Fistulas

*Jason S. Goldberg, Cristian Gragnaniello, Anthony J. Caputy, and Donald C. Shields*

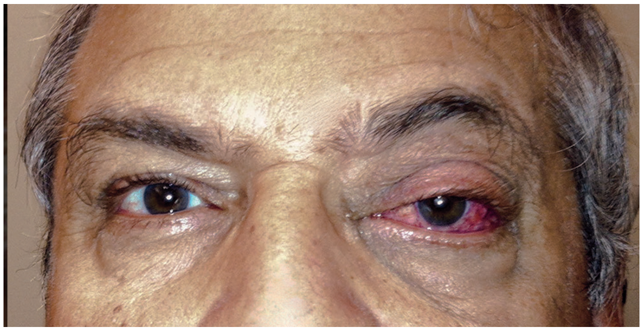

**Fig. 35.1** A 53-year-old male with left eye proptosis and conjunctival chemosis. (Source: Clinical Presentation. In: Spetzler R, Kalani M, Nakaji P, ed. Neurovascular Surgery. 2nd Edition. Thieme; 2015.)

## Clinical Presentation

- A 53-year-old male without significant past medical history presents to the emergency department following a closed head injury in a motor vehicle accident.
- He sustained a concussion, and the initial head CT scan was read as negative by the radiologist; he was admitted to the ICU due to somnolence, which improved over the next 12 hours.
- On day 2 in hospital, he was alert and oriented to person, place, and time, but he complained of worsening headache, diplopia, and an unusual noise with each heartbeat in the left side of his head.
- On day 3, the admitting physician noted left eye proptosis and conjunctival chemosis (▶**Fig. 35.1**). The patient complained of pain posterior to the left eye.

## Questions

1. Given the new symptoms and physical exam findings, what is the best next step in the management of this patient?
2. What is the most likely diagnosis, given the above data?
3. What information would you like to obtain from an ophthalmology consult?
4. You decide to reevaluate the initial head CT scan; what specific finding may be present?
5. What other common symptoms may this patient exhibit in addition to proptosis and conjunctival chemosis?
6. Which cranial nerve would you expect to be affected?
7. Describe the major treatment options and the potential complications associated with the most commonly utilized treatment modality.
8. How are carotid cavernous sinus fistulas (CCFs) classified, and what is this patient's likely prognosis?
9. What other mechanism, besides trauma, is a common cause of direct CCF and what features might predispose a patient to this cause?
10. What should patients expect regarding the resolution of their symptoms immediately following endovascular treatment and thereafter?

## Answers

1. *Given the new symptoms and physical exam findings, what is the best next step in the management of this patient?*
   - A four-vessel cerebral angiogram was ordered which revealed contrast media drainage into the cavernous sinus and superior ophthalmic vein (▶**Fig. 35.2**).
2. *What is the most likely diagnosis, given the above data?*
   - In conjunction with the cerebral angiogram, findings of proptosis, conjunctival chemosis, headache, and pulsatile whooshing noise suggest a diagnosis of CCF.[1,2,3,4]
3. *What information would you like to obtain from an ophthalmology consult?*
   - Formal visual acuity examination, fundoscopic evaluation, and intraocular pressure measurements provide the treating physician information relevant to the risk of visual loss in the affected eye.
4. *You decide to reevaluate the initial head CT scan; what specific finding may be present?*
   - Since the CCF in this patient was traumatic, a basilar skull fracture may be present; the fractured bone edges may have torn the carotid artery wall.[2]
5. *What other common symptoms may this patient exhibit in addition to proptosis and conjunctival chemosis?*
   - Presenting symptoms, such as headache, diplopia, retro-orbital pain, ophthalmoplegia, blurred vision, decreased visual acuity, and orbital bruits, are also common with CCFs.[1,2,3,4]
6. *Which cranial nerve would you expect to be affected?*
   - Cranial nerve VI is more likely to be affected due to its more medial course inferior to the carotid artery.
   - Cranial nerves III and IV are less often involved due to their course along the lateral wall of the cavernous sinus.[1]
7. *Describe the major treatment options and the potential complications associated with the most commonly utilized treatment modality.*
   - Treatment goal consists of preventing the fistulous drainage from the carotid artery into the cavernous sinus, while maintaining flow through the artery.[1]
   - Intracranial carotid artery ligation is rarely performed nowadays as endovascular techniques have evolved in recent decades.[2,3]
   - Transarterial or transvenous embolizations are commonly employed if conservative therapy results in worsening symptoms.[1,2,3]
   - Radiosurgery has been attempted in patients with indirect low-flow fistulas, but obliteration of the fistula may require months or years using this modality.[2]
   - Open surgical approaches may be employed if endovascular therapy is unsuccessful. Craniotomies for clipping, suturing, trapping, or sealing the fistula have been described.[1,2,5]
   - Complications associated with endovascular treatments include: retroperitoneal hematomas, ophthalmoplegias, cerebral infarcts, cavernous sinus rupture, cranial nerve palsies, intracranial (intracerebral and/or subarachnoid) hemorrhages, and extravascular extravasation of contrast.[1,2,3]
8. *How are CCFs classified, and what is this patient's likely prognosis?*
   - There are both formal and informal means of classifying CCFs.
     - Informally, they can be classified hemodynamically, anatomically, or by etiology.
       - Hemodynamically, the CCF can be high-flow or low-flow.
       - Anatomically, the CCF can be *direct* or *indirect*. A direct CCF is one that originates directly from the internal carotid artery (ICA) while an indirect CCF is one that originates from one of the branch vessels of the ICA.
       - Etiologically, the CCF can be *traumatic* or *spontaneous*.
     - Formally, one may use the *Barrow classification* of CCFs.[2,6] This system classifies four distinct types of CCFs using a type A through D nomenclature.
       - Type A is a direct, high-flow lesion resulting from a tear in the wall of the ICA. This is overwhelmingly the most common type of CCF (approximately 75–80% overall) and is secondary to trauma or aneurysm rupture.
       - Type B consists of indirect, low-flow lesions arising from the meningeal branches of the ICA.
       - *Type C* consists of indirect, low-flow lesions arising from the meningeal branches of the external carotid artery (ECA).
       - *Type D* CCF is an indirect, low-flow lesion arising from meningeal branches of *both* the ICA and ECA.
     - Overall prognosis for a CCF is favorable; more than 80% of patients receiving endovascular treatment achieve complete cure.[2,7] In clinically stable type B CCFs, a significant number of patients, as high as 90% in some series, may experience spontaneous resolution of the CCF over time.[8] Moreover, type D CCFs that have been converted to type B with endovascular treatment often resolve spontaneously.[9]

## Answers (*continued*)

9. *What other mechanism besides trauma, is a common cause of direct CCFs and what features might predispose a patient to this cause?*
   - Ruptured ICA aneurysms are a cause of direct, spontaneous CCFs. The demographic is typically an older, female patient.
   - Genetic syndromes that weaken the arterial wall, such as Ehlers–Danlos syndrome, pseudoxanthoma elasticum, and fibromuscular dysplasia, can predispose a patient to such spontaneous ruptures, which are often secondary to minor stresses.[1,2,3]

10. *What should patients expect regarding the resolution of their symptoms immediately following an embolization procedure and thereafter?*
    - Forty percent or more patients experience a transient worsening of their symptoms immediately following endovascular treatment. This usually resolves over time.
    - Features of CCFs, such as chemosis and proptosis, will resolve over hours to days, while cranial nerve palsies typically improve over the course of weeks.
    - Recovery of visual acuity is quite variable and will depend on cause, severity, and duration of loss.[1,2]

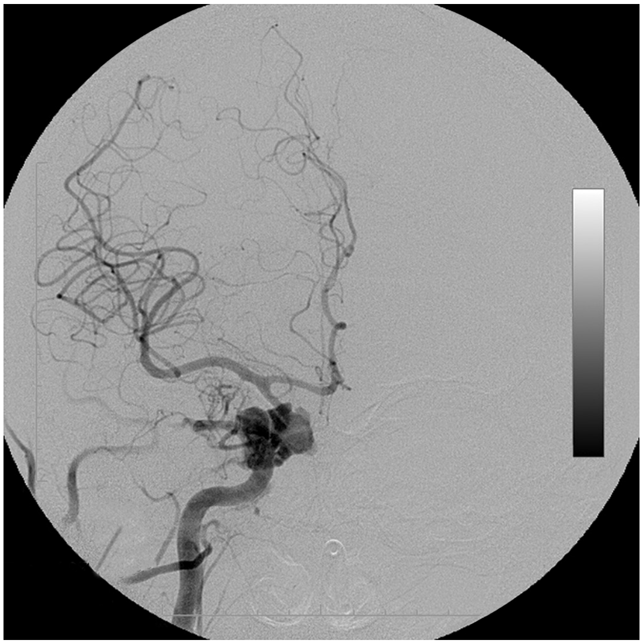

**Fig. 35.2** A four-vessel cerebral angiogram with anteroposterior view of right internal carotid artery selective injection reveals contrast in the cavernous sinus and the superior ophthalmic vein.

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# Case 36 Subarachnoid Hemorrhage and Vasospasm

*Qasim Al Hinai, Claude-Edouard Châtillon, David Sinclair, and Denis Sirhan*

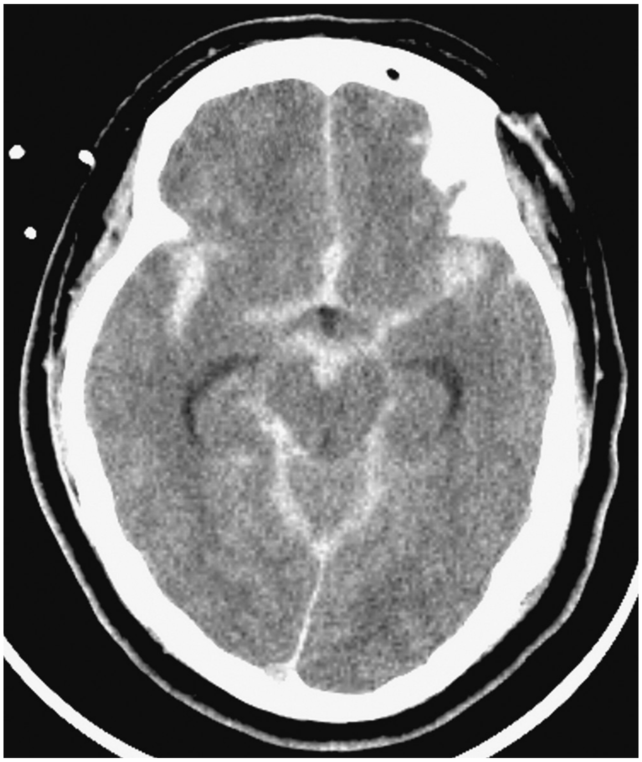

**Fig. 36.1** Noncontrast CT scan of the head, axial cut through basal cisterns.

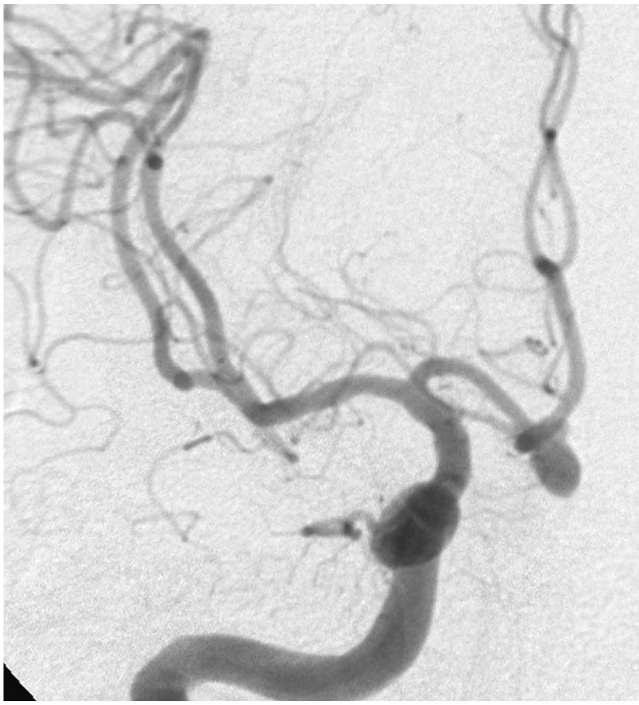

**Fig. 36.2** Cerebral angiogram, anteroposterior view, right carotid injection.

## Clinical Presentation

- A 55-year-old woman presents to the emergency room (ER) with sudden-onset severe headache, vomiting, and photophobia.
- On examination, she is confused and drowsy. There are no focal neurologic deficits. She opens her eyes spontaneously and obeys commands.
- CT scan of the head is obtained in the ER (▶**Fig. 36.1**).

## Questions

1. Interpret the CT scan (▶**Fig. 36.1**).
2. What is the diagnosis?
3. Describe two clinical grading scores of your diagnosis. What is the grade in this case?
4. Give one radiologic grading system of your diagnosis and its prognostic significance. What is the grade in this case?
5. What is your management?
6. Describe the cerebral angiogram shown in ▶**Fig. 36.2**.

   The patient was admitted to the intensive care unit (ICU) and underwent a therapeutic procedure for the findings on the previous angiogram. Six days later, the patient developed left-sided hemiparesis (left leg is weaker than left arm). A CT scan of the head revealed a right mesial frontal hypodensity with surrounding mild edema.

7. What is the most likely diagnosis and what studies do you obtain?
8. Describe the cerebral angiogram shown in ▶**Fig. 36.3**.
9. What is the pathophysiology of this condition?
10. What additional investigations will help you in the management at this stage?

## Questions (*continued*)

11. What intervention(s) are possible during angiography?
12. How will you treat this condition medically?

Patient improved with the above treatment except for mild left-sided hemiparesis. She was discharged home. Four weeks later, she presented to the ER with moderate to severe headache. Plain CT scan of head is shown (▶**Fig. 36.4**).

13. Interpret the CT scan shown in ▶**Fig. 36.4**.
14. What is the management?

## Answers

1. *Interpret the CT scan (▶***Fig. 36.1***).*
   - There is an extensive subarachnoid hemorrhage (SAH) involving the anterior interhemispheric fissure, bilateral Sylvian fissures, and interpeduncular and crural cisterns. The temporal horns of the lateral ventricles are dilated.
2. *What is the diagnosis?*
   - SAH, most likely secondary to aneurysmal rupture.
3. *Describe two clinical grading scores of your diagnosis. What is the grade in this case?*
   - The two most commonly used grading systems are Hunt and Hess (H&H) and World Federation of Neurosurgical Societies (WFNS) grading scores.
     - Hunt and Hess grading:[1]
       - 1: Asymptomatic or mild headache (H/A) and slight nuchal rigidity
       - 2: Cranial nerve palsy, moderate to severe H/A, and nuchal rigidity
       - 3: Mild focal deficit, lethargy, or confusion
       - 4: Stupor, moderate to severe hemiparesis, and early decerebrate rigidity
       - 5: Deep coma, decerebrate rigidity, and moribund appearance
     - WFNS grading[2]:
       - 1: Glasgow Coma Score (GCS) 15 with no major focal deficit
       - 2: GCS 13 to 14 with no major focal deficit
       - 3: GCS 13 to 14 with major focal deficit
       - 4: GCS 7 to 12 with or without major focal deficit
       - 5: GCS 3 to 6 with or without major focal deficit
   - This patient has an H&H grade 3 and WFNS grade 2.
4. *Give one radiologic grading system of your diagnosis and its prognostic significance. What is the grade in this case?*
   - Modified Fisher scale[3]:
     - 0: No SAH or IVH
     - 1: Focal or diffuse thin SAH, no IVH
     - 2: Focal or diffuse thin SAH, with IVH
     - 3: Thick SAH present, no IVH
     - 4: Thick SAH present, with IVH
   - Modified Fisher scale is more sensitive in predicting the outcome. This pt has modified Fisher scale 3.
   - The amount of subarachnoid blood correlates with the risk of vasospasm.
   - Scale 4 carries the worst prognosis.
5. *What is your management?*

   Please refer to Case 44 [Stent and balloon assisted coiling] for a systemic approach to this answer.
   - Management includes the following steps:[4]
     - ICU admission
     - Arterial and central venous lines. Monitoring volume status in certain patients with recent aneurysmal SAH (aSAH) by central venous pressure (CVP) is reasonable, as is treatment of volume contraction with crystalloid or colloid fluids *(Class IIa; Level of Evidence B)*[5].
     - Monitor systolic blood pressure (SBP) and mean arterial pressure (MAP). Between the time of aSAH symptom onset and aneurysm obliteration, blood pressure should be controlled with a titratable agent to balance the risk of stroke and hypertension-related rebleeding, and maintain cerebral perfusion pressure *(Class I; Level of Evidence B)*[5]. The magnitude of blood pressure control to reduce the risk of rebleeding has not been established, but a decrease in SBP to < 160 mm Hg is reasonable *(Class IIa; Level of Evidence C)*[5].
     - Administer phenytoin (Dilantin) loading (18 mg/kg) and maintenance doses (100 mg tid × 1 week unless seizures). Retrospective studies identified risk factors for the development of early seizures associated with aSAH, including aneurysm in the middle cerebral artery, thickness of aSAH clot, associated intracerebral hematoma, rebleeding, infarction, poor neurological grade, and history of hypertension.[5] The use of prophylactic anticonvulsants may be considered in the immediate posthemorrhagic period *(Class IIb; Level of Evidence B)*.[5]
     - Administer nimodipine (60 mg by mouth q4h × 21 days). The administration of nimodipine to reduce the risk of poor outcome and delayed cerebral ischemia (DCI) is the only level IA evidence recommended by the American Stroke Association (ASA) *(Class I; Level of Evidence A)*[5].
     - Analgesics
     - In cases of hydrocephalus or H&H grade ≥ 3, an external ventricular drain (EVD) should be inserted. This enables accurate intracranial pressure (ICP) measurement along with adequate CPP management (>60 mm Hg). Aneurysmal

## Answers (*continued*)

SAH-associated acute symptomatic hydrocephalus should be managed by cerebrospinal fluid diversion (EVD or lumbar drainage, depending on the clinical scenario) *(Class I; Level of Evidence B)*[5].

- Cerebral angiography +/- coiling (if the cause of SAH is aneurysm). CT angiography (CTA) or Digital subtraction angiography (DSA) with three-dimensional rotational angiography is indicated for both detection of aneurysm(s) in patients with aSAH and for planning treatment (to determine whether an aneurysm is amenable to coiling or to expedite microsurgery) (Class I; Level of Evidence B).[5,6] If the aneurysm is not reasonably coilable, then craniotomy and aneurysm clipping should be entertained.
- In cases of poor H&H grade (IV and V) at presentation, prognosis is very poor unless their low GCS is partly reversible due to hydrocephalus or to a postictal state. Initial management consists of an EVD insertion, maximizing CPP and observation for a few hours.

6. *Describe the cerebral angiogram shown in* ▶ **Fig. 36.2**.
The right anteroposterior projection right internal carotid artery angiogram reveals an aneurysm at the level of the anterior communicating artery (ACOM) pointing inferiorly.
7. *What is the most likely diagnosis and what studies do you obtain?*
   - The likely diagnosis is severe radiological vasospasm affecting the right anterior cerebral artery (ACA), and mild to moderate angiographic vasospasm affecting the right intracranial ICA, and right MCA branches.
   - Vasospasm is a major complication of SAH.
   - Clinical vasospasm occurs in 30% cases of SAH.
   - Infarction occurs in about half of these cases if vasospasm remains untreated.
   - Seven percent of attributed deaths are due to vasospasm.[7]
   - Very few preventive and therapeutic measures have reproducible benefits in randomized trials.
   - A cerebral angiogram may be done to confirm the presence of radiographic vasospasm, which may correlate with the clinical picture. Vasospasm after SAH is best evaluated by DSA or high-definition CT angiography.[8]
8. *Describe the cerebral angiogram shown in* ▶ **Fig. 36.3**.
   - As suspected clinically, the angiogram reveals severe narrowing suggesting cerebral vasospasm affecting the right A2 segment of the ACA.
   - Note the occluded right ACOM aneurysm which has been previously treated by endovascular coiling.
9. *What is the pathophysiology of this condition?*
The exact pathophysiology is unknown. However, there are theories that might explain vasospasm pathophysiology[9]:
   - Theories of spasmogens
     - Components of erythrocytes
     - Oxyhemoglobin
     - Plasma and white blood cells appear to not induce vasospasm
   - Structural theories
     - Proliferative vasculopathy
     - Immune vasculopathy
     - Vessel wall inflammation
     - Extracellular lattice contraction
   - Vasoconstriction theories
     - Free radical lipid peroxidation
     - Derangement in eicosanoid production
     - Nitric oxide deficit
     - Endothelin excess[4]
10. *What additional investigations will help you in the management at this stage?*
    - Transcranial Doppler (TCD): TCD examinations are noninvasive, carried out at the bedside, and can easily be performed on a daily basis providing patients have an adequate acoustic window in their temporal region through which to insonate.[10] TCD is reasonable to monitor for the development of arterial vasospasm *(Class IIa; Level of Evidence B)*.[5] Impaired cerebral autoregulation assessed by TCD was found to be predictive of vasospasm when detected within the first few days following SAH in a recent single-center study.[11]
    - Electroencephalogram (EEG)
    - Cerebral blood flow (CBF) studies using positron emission tomography (PET), single-photon emission CT (SPECT), xenon-enhanced CT, perfusion CT, thermal diffusion flowmetry, and diffusion-weighted MRI scans.[10] Perfusion imaging with CT or MR can be useful to identify regions of potential brain ischemia *(Class IIa; Level of Evidence B)*.[5] In a meta-analysis, the sensitivity of perfusion CT scan in identifying patients with angiographic vasospasm was 74% and specificity was 93%.[12]
11. *What intervention(s) are possible during angiography?*
Vasodilation by balloon angioplasty (if focal vasospasm present) and/or selective intra-arterial milrinone, nifedipine, nimodipine, verapamil, and/or papaverine (in the case of diffuse Circle of Willis (COW) spasm) is reasonable in patients with medically refractory symptomatic cerebral vasospasm (i.e. those who are not rapidly responding to hypertensive therapy) (Class IIa; Level of Evidence B).[5,10]
12. *How will you treat this condition medically?*
    - Induced hypertension10: Keep SBP at 160 mm Hg or MAP at 120 mm Hg or above. If an ICP monitor is in situ, maintain CPP > 60 mm Hg.
    - Triple H ( hypertension, hypervolemia and hemodilution ) therapy in the treatment of cerebral vasospasm has never been properly assessed

## Answers (*continued*)

for efficacy in a control trial.[13] Hypervolemia can increase the risk of cardiorespiratory complications. Ensure euvolemia with strict fluid balance. (CVP, if measured, is kept around 6 mm Hg).

- The focus shifted from triple-H therapy to the maintenance of euvolemia and induced hypertension.[10]
- Normonatremia: [Na] > 140 mEq/L
- Normoglycemia: [glucose] < 8 mmol/L. Careful glucose management with strict avoidance of hypoglycemia may be considered as part of the general critical care management of patients with aSAH *(Class IIb; Level of Evidence B)*.[5]
- Normothermia: aggressive control of fever to a target of normothermia by use of standard or advanced temperature-modulating systems is reasonable in the acute phase of aSAH *(Class IIa; Level of Evidence B)*.[5]
- Norepinephrine bitartrate (Levophed; Abbott Laboratories, Abbott Park, IL): It may be used to maintain blood pressure at pretreatment levels on an as needed basis. Provided that the ruptured aneurysm has been repaired, symptomatic vasospasm should be treated by the administration of a vasopressor, the most commonly used being phenylephrine or norepinephrine.[14]
- Milrinone: A phosphodiesterase 3 inhibitor appears to be a promising agent in reducing the incidence of vasospasm may also be used in an intravenous infusion form. Recently, the Montreal Neurological Hospital protocol has been reported.[15] An intravenous milrinone infusion was used for a mean of 10 days without any significant side effects. No medical complications associated with this protocol were observed. There were five deaths; of the surviving patients, 48.9% were able to go back to their previous baseline and 75% had a good functional outcome.[15]

13. *Interpret the CT scan shown in* ▸**Fig. 36.4**.
    - The CT scan shows obliteration of cerebral convexity sulci and dilation of all ventricles indicating the development of chronic, SAH-associated communicating hydrocephalus.
14. *What is the management?*
    - Insertion of ventriculoperitoneal shunt. Aneurysmal SAH-associated chronic symptomatic hydrocephalus should be treated with permanent cerebrospinal fluid diversion *(Class I; Level of Evidence C)*.[5]

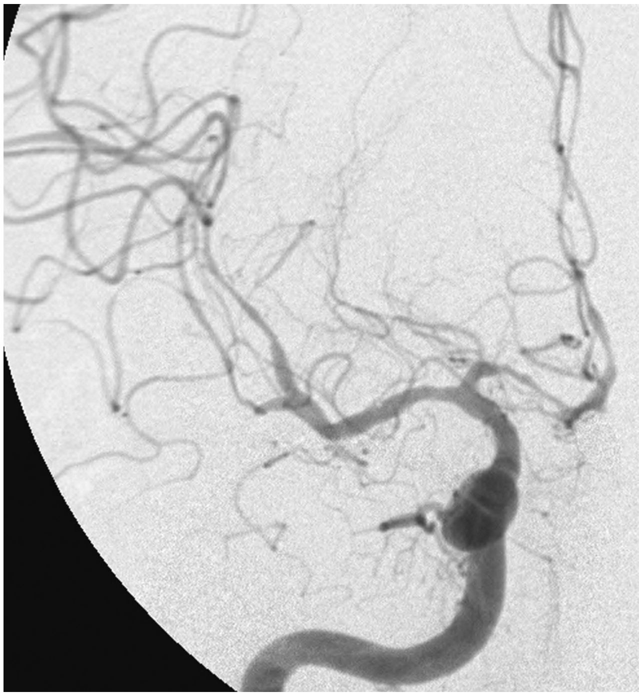

**Fig. 36.3** Cerebral angiogram, anteroposterior view, right carotid injection, performed 6 days later, after a therapeutic intervention.

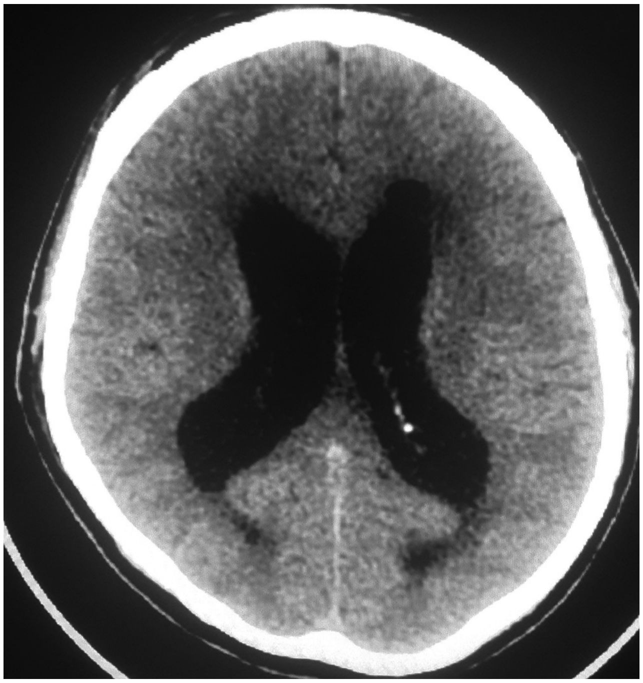

**Fig. 36.4** A plain CT scan of head, axial cut through lateral ventricles.

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# Case 37 Posterior Communicating Artery Aneurysm

*Badih Daou, Pascal M. Jabbour, and Erol Veznedaroglu*

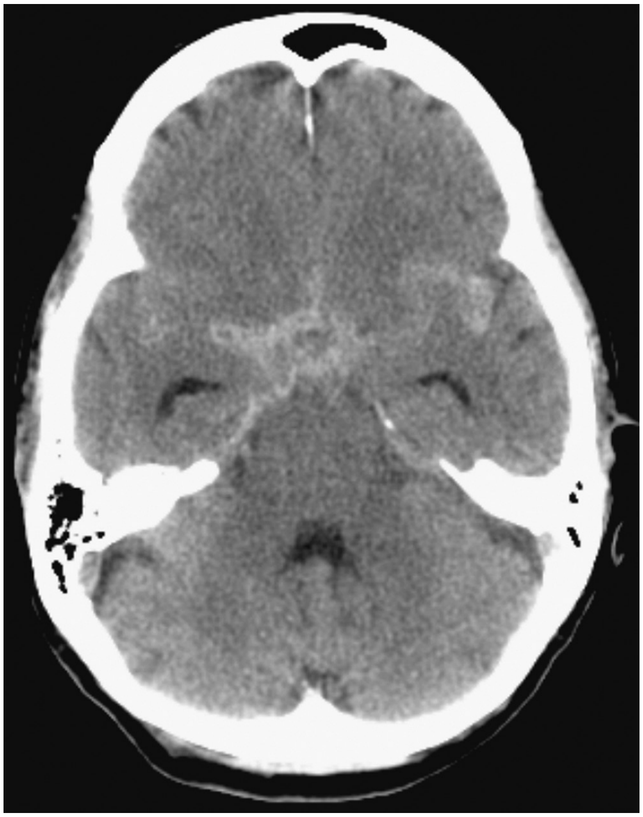

**Fig. 37.1** CT scan of the head showing diffuse subarachnoid hemorrhage.

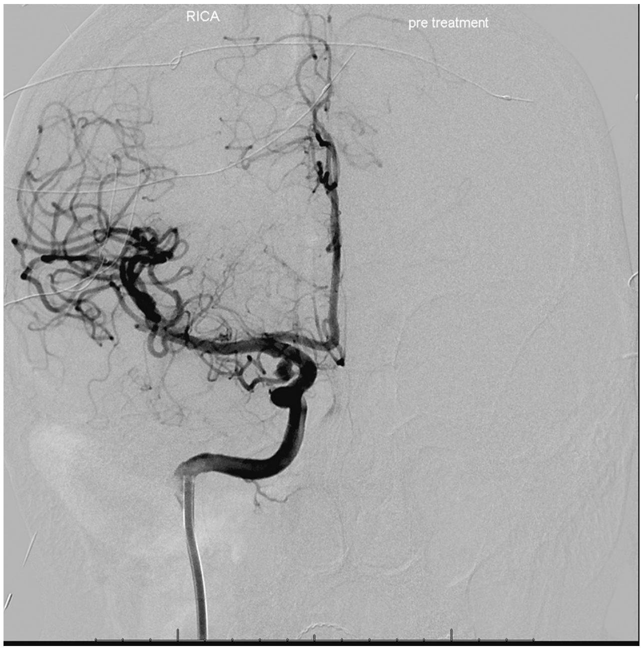

**Fig. 37.2** Cerebral angiogram, right carotid injection showing a cerebral aneurysm.

## Clinical Presentation

- A 64-year-old right-handed woman with a history of high blood pressure and smoking presents to the emergency room after experiencing the worst headache of her life while she was driving her car.
- She does not have a history of migraine headaches and never complained of headaches before this episode.
- Her familial history is significant—a maternal aunt died 10 years ago from a ruptured brain aneurysm.
- Neurologic evaluation showed that the patient was alert, awake, oriented, with a dilated nonreactive pupil on the right side with decreased ocular motility upward, inward and downward.
- A CT scan of the head is shown in ▶**Fig. 37.1** and a cerebral angiogram is shown in ▶**Fig. 37.2**.

## Questions

1. What is the most likely diagnosis based on clinical history?
2. What are this patient's Hunt and Hess and Fisher grades?
3. What are the risk factors for subarachnoid hemorrhage (SAH) in this patient?
4. How are the findings on the clinical examination of this patient relevant; and where is the most likely anatomical location of the lesion based on the imaging studies provided?
5. What are some common characteristics of this type of aneurysms?
6. What are the treatment options available for this patient?
7. You opt to proceed with surgical repair. Describe the details of the operation including positioning, opening, details of dissection and repair, closure, and other assistive measures.

## Answers

1. *What is the most likely diagnosis based on clinical history?*
   - The patient experienced the worst headache of her life, with no previous history of headaches. This is highly suspicious of SAH.
2. *What are this patient's Hunt and Hess and Fisher grades?*
   - The patient has a Hunt and Hess grade 2 and a Fisher grade 3.[1,2]
3. *What are the risk factors for subarachnoid hemorrhage (SAH) in this patient?*
   - Female, high blood pressure, smoker, and familial history of aneurysms[3]
4. *How are the findings on the clinical examination of this patient relevant; and where is the most likely anatomical location of the lesion based on the imaging studies provided?*
   - The patient has a partial third nerve palsy involving the pupil.
   - Most probably the patient has a right posterior communicating artery (PCOM) aneurysm because of the proximity of the third nerve to the PCOM.
5. *What are some common characteristics of this type of aneurysms?*
   - PCOM aneurysms are the second most common aneurysms overall (25% of all aneurysms) representing 50% of all internal carotid artery (ICA) aneurysms.[4]
   - Although patients with a PCOM aneurysm typically present with SAH, they can also present with subdural hematoma or isolated oculomotor nerve palsy. About 20% of PCOM aneurysms have third nerve palsy on presentation which is usually secondary to some conformational change in the dome of the aneurysm and often indicates an impending rupture.[4]
6. *What are the treatment options available for this patient?*
   - The patient's treatment options are either open surgery and clip ligation of the aneurysm or endovascular treatment with coils with or without stent placement, depending on the patient's comorbidities, the shape of the aneurysm and the neck, and the preference of the surgeon. Coiling of PCOM aneurysms is associated with lower morbidity; however, coiled PCOM aneurysms have a higher risk of recurrence and must be followed closely.[5]
   - Surgical clipping may be favored in PCOM aneurysms associated with a fetal posterior communicating artery. A fetal PCOM variant is defined as a PCOM artery, which has the same caliber as the P2 segment of the posterior cerebral artery (PCA) and is associated with an atrophic P1 segment. Because fetal PCOM arteries are the primary supply to the PCA, clipping is favored over endovascular therapy to minimize the risk of compromising flow to this artery.[4]
   - Both surgical clipping and embolization are safe and effective methods resulting in functional third nerve recovery. Studies that directly compared the two treatment modalities concluded that clipping offers a higher chance of oculomotor nerve palsy recovery; however, coiling will remain an option particularly in elderly patients or patients with significant comorbidity.[6,7]
   - Factors associated with a higher likelihood of recovery include earlier time to treatment, partial third nerve deficit, and presence of SAH.[4]
7. *You opt to proceed with surgical repair. Describe the details of the operation including positioning, opening, details of dissection and repair, closure, and other assistive measures.*
   - Positioning and preoperative preparation
     - Supine, shoulder roll, head rotated 45 degrees, Mayfield 3-point fixation
     - Preoperative antibiotics, mannitol, furosemide available
     - Phenobarbital or etomidate available
     - Ventriculostomy ready to be placed
     - Assistive devices: microscope, loops, headlight, ultrasound; if have intraoperative angiography, have it ready

## Answers (*continued*)

  - Retractors: Greenberg, Fukushima; Lela bar or Budde halo
  - Consider somatosensory evoked potential, electroencephalogram intraoperatively, if feasible.
  - Anesthetize the pin sites before pinning.
  - Prepare the neck for possible early proximal control.
- Opening and dissection
  - Curvilinear incision, pterional craniotomy
  - Take down sphenoid wing extradurally to the meningo-orbital artery.
  - Clinoid may have to be partially removed to obtain adequate exposure and proximal control of the internal carotid artery.
  - Wax all bone edges to prevent air embolism.
  - Place tack-up sutures.
  - Open the dura based on sphenoid wing.
  - Split the fissure under the microscope from proximal to distal.
  - Open between the veins and frontal cortex.
  - You may place retractors on frontal lobe and gently start sharp dissection.
  - Identify the optic nerve and carotid artery and dissect the carotid to be able to place a proximal clip.
  - Work your way back to identify the carotid bifurcation.
  - Wide opening of the Sylvian fissure greatly facilitates the safety of clipping.
  - Dissection should be done with caution in laterally projecting aneurysms to avoid avulsion of the fundus from the temporal lobe attachments.
  - Approach directed more frontal is preferred until the aneurysm neck is visualized.
- Clipping and precautions
  - You may then place temporary clips and work on dissecting the neck and dome.
  - Note: Internal carotid artery can be clipped up to 15 minutes before opening and reperfusing the brain.
  - Use systemic hypertension, phenobarbital or etomidate, mild hypothermia, mannitol during temporary clip.
  - Dissect neck before the dome, then place clip, then dissect dome and check for perforators.
  - Advance the clip just beyond the course of the PCOM, without compromising the patency of that artery or that of the anterior thalamoperforators, internal carotid perforators, or anterior choroidal artery.
  - Use ultrasound to the dome to verify that there is no flow.
  - If there is no flow and no perforators, open the dome.
  - Open the membrane of Liliequist widely to visualize and release any tethering or compromise of the PCOM and its thalamoperforating vessels.
  - Obtain intraoperative angiogram, if possible, to confirm patency.
- Closure
  - Close and transfer the patient to intensive care unit.
  - Obtain immediate postoperative CT scan and angiogram as well as neurologic exam.

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# Case 38 Middle Cerebral Artery Aneurysm with Intracerebral Hemorrhage

*Nicholas J. Erickson, Cristian Gragnaniello, Marguerite Harding, Zachary N. Litvack, Anthony J. Caputy, Remi Nader, and Dimitri Sigounas*

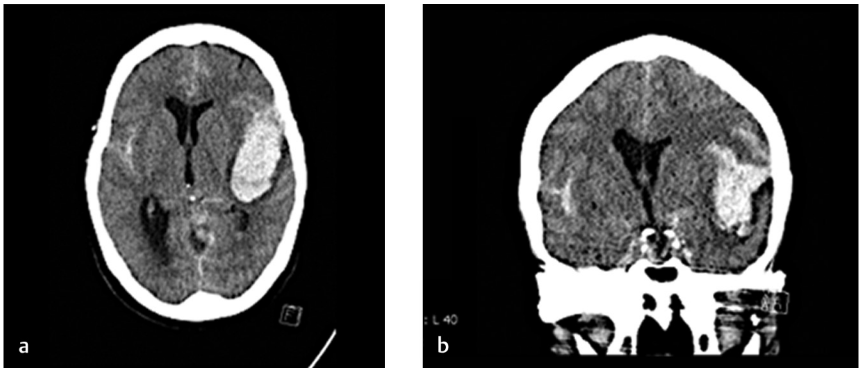

**Fig. 38.1** Initial CT. Axial (**a**) and coronal (**b**) views demonstrating a subarachnoid hemorrhage with intracerebral hemorrhage with extension into the left frontotemporal region.

## Clinical Presentation

- A 62-year-old man presents to the emergency department with sudden onset of severe headache and stiff neck. He states that symptoms started 2 hours ago. He has also vomited four times in the past 3 hours and complains of difficulty in speaking as well as right-sided upper and lower extremity weakness. The patient has a history of hyperlipidemia, hypertension, and has smoked a pack per day for the past 30 years. Initial CT scan is performed (▶**Fig. 38.1**).
- While he is being assessed, the patient's neurologic status declines and he loses consciousness, falls to the floor, and has a witnessed generalized seizure. The patient is intubated, stabilized, and a CT angiogram (CTA) of head is performed (▶**Fig. 38.2**) along with a second CT scan.

## Questions

1. Describe your diagnosis.
2. What are the next steps in the management of this patient?
3. What is your definitive choice of treatment and why?
4. How is subarachnoid hemorrhage (SAH) graded? What other factors are to be considered in this case?
5. Describe a surgical strategy for this case.
6. Describe the role of endovascular treatment.
7. What are the late complications of middle cerebral artery (MCA) aneurysm rupture with intracerebral hemorrhage (ICH)?
8. What are the rates of morbidity, mortality, and residuals based on treatment modality in this case and in cases of unruptured aneurysms in the same location?

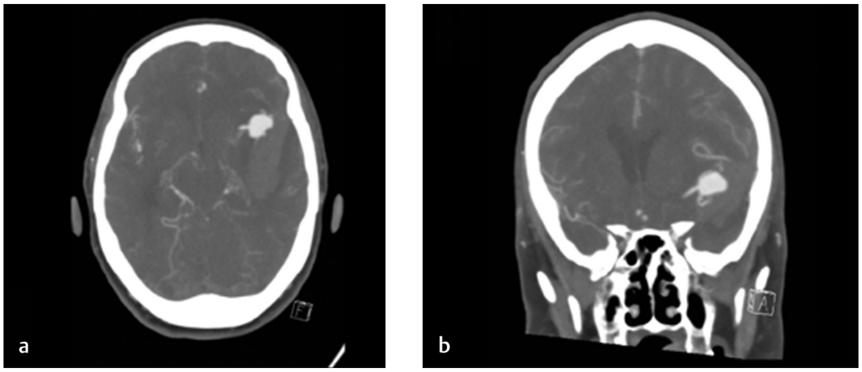

**Fig. 38.2** CT angiography. Axial (**a**) and coronal (**b**) views showing a large middle cerebral artery aneurysm with extravasation into a hematoma.

## Answers

1. *Describe your diagnosis.*
   - The clinical and imaging findings demonstrate an SAH with ICH extending into the left frontotemporal region.
   - On axial and coronal CT images, the left Sylvian fissure appears to be expanded by an enlarging hematoma, causing associated midline shift. The sagittal CT reconstruction shows diffuse SAH tracking out of the Sylvian fissure. This distribution of blood is most consistent with a ruptured left MCA aneurysm.
   - CTA shows a large MCA aneurysm with extravasation into a hematoma.
2. *What are the next steps in the management of this patient?*
   - Based on the clinical findings, the initial management of this patient should include:
     - Rapid resuscitation
     - Immediate intracranial pressure (ICP) control: reduction of ICP can be initially achieved with mannitol (1 g/kg)
     - Noncontrast head CT and CTA
     - Basic laboratory studies to asses for hypocoagulability, platelet count, and sodium level (complete blood count [CBC], electrolytes, prothrombin time [PT], partial thromboplastin time [PTT]).
     - The intracerebral hematoma should be immediately evacuated surgically as it causes symptomatic mass effect on the brain.
     - If SAH is due to aneurysmal rupture, early aneurysmal occlusion/repair within 24 hours should be performed.
     - Prophylaxis against delayed cerebral ischemia should be planned.
     - The patient should be cared for in a neuro intensive care unit (ICU) with continued monitoring of ICP, hypervolemia, and liberalized blood pressure parameters after aneurysmal obliteration.
     - This patient requires immediate intubation.
     - Intravenous (IV) access should be established via central line placement.
     - Seizure control with diazepam and stat dosing of phenytoin.
   - Anticoagulation history in this patient is important as most experts favor reversal of all anticoagulants until the aneurysm is definitively repaired by surgery or coiling in addition to maintaining normotension. [1]
3. *What is your definitive choice of treatment and why?*
   - Emergent surgical treatment is the definitive choice. This involves an immediate craniotomy with a large bone flap in the event of brain herniation and the need to leave the flap off, evacuation of the hematoma and surgical clipping of the aneurysm as well as reconstruction of the parent vessel.
   - MCA aneurysms most commonly arise from the proximal bifurcation of the MCA trunk near the M1–M2 junction. Occasionally, they will arise at the origin of the anterior temporal artery, lateral lenticulostriate arteries, secondary bifurcation or more distally.
   - The best proven method to treat intracerebral hematoma due to a ruptured MCA aneurysm is by direct hematoma evacuation and microsurgical clipping. The superficial location, frequent broad-based aneurysmal neck configuration, and relatively straightforward surgical anatomy make these aneurysms more amenable to surgical clipping than to endovascular coiling.[2,3]
4. *How is subarachnoid hemorrhage (SAH) graded? What other factors are to be considered in this case?*
   - The Hunt and Hess (H&H) scale is used to classify the severity of a subarachnoid hemorrhage and is based on the patient's clinical condition. Admission H&H grade is the primary prognostic factor for outcome in this patient population. Grading ranges from I to V and takes into account clinical findings such as headache, nuchal rigidity, focal neurologic deficits, mental status, and hemiparesis. Studies suggest patients presenting with grade IV or V SAH may not benefit from aggressive treatment.
   - The World Federation of Neurosurgical Societies (WFNS) scale uses the Glasgow Coma Scale combined with the presence or absence of focal deficits to determine the severity of SAH and to predict patient outcomes. This scale is now used more widely than the H&H scale.

## Answers (*continued*)

- Other prognostic factors include:
  - Timing: many studies have shown that the time to treatment is a very important factor, even for poor grade patients. Early surgery within 6 to 12 hours of hemorrhage has shown the most promising outcomes.
  - Other findings, such as young age, better clinical grade, and small ICH volume (< 25 mL) are associated with better outcomes. Shimoda et al noted patients with ICH and diffuse SAH fared worse compared to those with aneurysm associated with only ICH, due to the increased risk of vasospasm in the former patients.[3]

5. *Describe a surgical strategy for this case.*
   - The primary goal of the surgical procedure contemplated initially should be the evacuation of the hematoma, and addressing the brain edema/ increased ICP and mass effect. If the aneurysm is not safely attainable during the initial surgical procedure, the surgeon should strongly consider aborting the aneurysm repair portion of the surgery and should come back at a later time once the edema is under control or, alternatively, consider an endovascular approach to securing the aneurysm.
   - A large, left pterional craniotomy or a large left-sided trauma flap is appropriate for this case to prevent brain herniation and incarceration as well as to facilitate access to the hemorrhage (see ▶**Fig. 38.3** and ▶**Fig. 38.4** for intraoperative image and postoperative CT findings in this case).
   - Pertinent surgical points:
     - Patient head positioning, extent of bony exposure, decision to place a ventriculostomy, and evacuation of hematoma should all be part of operative planning.
     - The patient's head, fixed into a Mayfield head holder should be turned to the right side 45 degrees relative to the floor with the neck slightly extended making the malar eminence the highest point in the surgical field.
     - Large pterional craniotomy with drilling of the lesser wing of the sphenoid is performed. Also, one should try to extend the bony opening as low as possible toward the floor of the middle fossa to avoid damage to the parenchyma which may be compressed against bony edges during dissection.
     - If possible, one should release cerebrospinal fluid (CSF) from the carotid cistern. Also, a ventriculostomy at Paine's point can be useful.
     - Wide dissection of the Sylvian fissure is performed to identify the parent and distal vessels as well as the aneurysm to permit for proximal and distal control with temporary clipping.
     - In most cases, evacuation of the ICH will facilitate Sylvian fissure dissection, despite the brain swelling, as this will provide a working corridor.
     - One should avoid excessive brain retraction/ manipulation. This leads to further postoperative brain swelling, risk of venous compression or infarction, and increased morbidity.
     - One should avoid blunt dissection around the aneurysm, as this can precipitate an intraoperative rupture.
     - Reconstruction of an MCA bifurcation aneurysm may prove difficult depending on the specific anatomy of the aneurysm and its base. On occasion, a simple reconstruction may not be feasible and reconstruction of the neck may require bypass.[4–6]
     - Topical application of vasodilators may help in preventing postoperative vasospasm.
     - The bone flap in some instances of significant edema has to be stored rather than replaced at the time of surgery and subsequently replaced 6 to 8 weeks postoperatively.
6. *Describe the role of endovascular treatment.*
   - The role of endovascular coiling in poor grade patients has only been described in limited clinical series; furthermore, there were very few poor grade patients included in the International Subarachnoid Aneurysm Trial (ISAT). Some studies suggest that endovascular treatment may be appropriate in some poor grade patients when combined with open therapies, especially in the presence of ICH. Suzuki et al showed favorable outcomes in 43.8% of H&H grade IV and 12.9% of H&H grade V patients with combined approaches to treatment.[2]
   - One potential advantage of endovascular treatment is that it does not require extensive retraction of brain tissue and vessel dissection. This is much less physiologically stressful for the patient and may be beneficial when extensive cerebral swelling is seen on CT scan.[2]
   - Despite recent advancements in endovascular coiling, clipping is still the preferred choice of treatment for MCA aneurysms. The common occurrence of these aneurysms at the bifurcation, incorporation of M2 divisions, along with their wide necks and association with expanding hematomas make them less suitable for coiling in general.[3–5]
   - ▶**Table 38.1** outlines favorable conditions for coiling versus clipping.[7]
7. *What are the late complications of middle cerebral artery (MCA) aneurysm rupture with intracerebral hemorrhage (ICH)?*
   - Outcomes for aneurysmal rupture depend on several factors. Historically, outcomes associated with SAH and ICH are associated with significant morbidity.
   - Early complications mainly include rebleeding and vasospasm.

## ■ Answers (*continued*)

- Late complications include:
  - Seizure: incidence is between 7 to 25% with one MCA aneurysm. Most patients with ruptured MCA aneurysms presenting with temporal ICH will develop delayed seizures.
  - Visual deficits: associated with lesions in the loop of Meyer.[4,7,8]
  - Weakness due to proximity to motor cortex
  - Dysphasia due to proximity to speech centers

8. *What are the rates of morbidity, mortality, and residuals based on treatment modality in this case and in cases of unruptured aneurysms in the same location?*
   - Overall, surgical morbidity rates are ~30% for ruptured aneurysms.[9]
   - Based on the ISAT and International Study of Unruptured Intracranial Aneurysms (ISUIA) studies, the outcomes are outlined in ▶**Table 38.2.**[4,9,10]
   - Other more recent studies have shown the following:
     - The 6-year results of the Barrow Ruptured Aneurysm Trial (BRAT) study were recently published and were based on the following: 471 patients were randomly assigned to either coiling or clipping treatments. For anterior circulation aneurysms, the study showed no significant difference in outcomes (death and dependency) between coiling and clipping but a persistent benefit was observed in the coiling group for treating posterior circulation aneurysms.[11]
     - Complete occlusion rates were observed to be clearly lower in the coiling group, and the results of the Cerebral Aneurysm Re-Rupture After Treatment (CARAT) study, showed a strong association between the degree of residual aneurysm size and risk for rehemorrhage.[12] For patients having an aneurysmal rupture after treatment, the mortality rate in the CARAT study was 58%.[12]
     - The Cerecyte coil trial showed a combined success rate, defined as stable angiographically visible occlusion, stable neck remnant, or improved occlusion of the aneurysm, in 57% cases (245/433) for ruptured and unruptured aneurysms at the 6-month follow-up mark after aneurysm treatment with Cerecyte coils.[13]
     - The hydrogel-coated coils versus bare platinum coils for the endovascular treatment of intracranial aneurysms (HELPS) trial reported significant angiographically confirmed recurrences in aneurysms treated using hydrogel-coated coils: 24 to 33% of patients had recurrences at the 18-month follow-up mark[14]. However, posttreatment (endovascular) hemorrhage rates were seen to be very low in the same group (0.2–1.3%).[14]
     - Overall, the best endovascular results, including those for treatment of ruptured and unruptured aneurysms, show complete obliteration in only less than 60% of the time.[11]

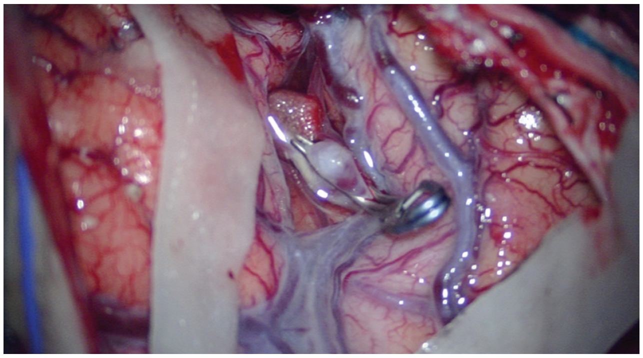

**Fig. 38.3** Clipping. Intraoperative view demonstrating successful exposure and clipping of the middle cerebral artery aneurysm.

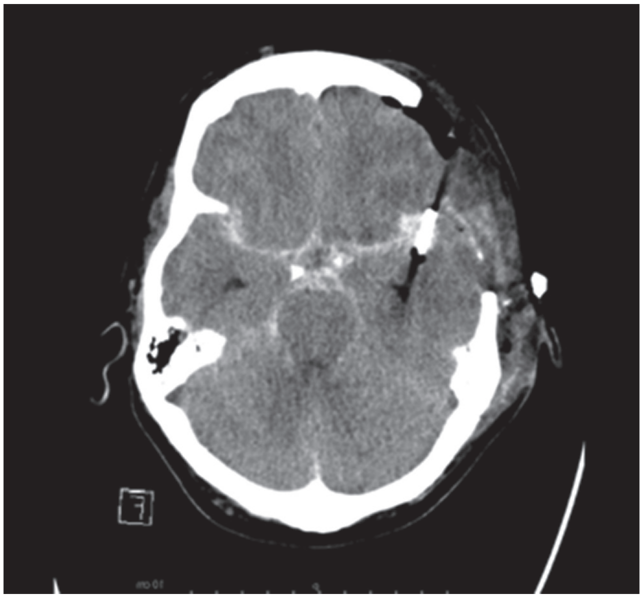

**Fig. 38.4** Postoperative CT scan. Axial view of noncontrast CT demonstrating successful evacuation of the hematoma and clipping of the aneurysm. The bone flap will be replaced 6 to 8 weeks postoperatively.

**Table 38.1** Conditions to consider conditions for coiling versus clipping

| Coiling | Clipping |
|---|---|
| Elderly (> 75 yr) | Younger age: lower risk of surgery and lower recurrence risk |
| Poor clinical grade | MCA bifurcation aneurysms |
| Inaccessible ruptured aneurysms | Giant aneurysms: > 20 mm diameter |
| Aneurysm configuration | Symptoms due to mass effect |
| A: dome-to-neck ratio ≥ 2 | Small aneurysms: < 1.5–2 mm diameter |
| B: absolute neck diameter < 5 mm | Wide aneurysm neck |
| Posterior circulation aneurysms | Patients with residual filling after coiling |
| Patients on Plavix | |

**Table 38.2** Morbidity and mortality rates based on treatment modality in cases of ruptured and unruptured aneurysms

| | | |
|---|---|---|
| Ruptured | Surgical | 30% |
| | Endovascular | 23% |
| | Observation (30-day mortality) | 45% |
| | Observation (serious neurologic deficit) | 25% |
| Unruptured | Surgical | 12% |
| | Endovascular | 9% |

*Source:* Data from Molyneux et al 2005; International Study of Unruptured Intracranial Aneurysms Investigators 1998.[9,10]

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# Case 39 Distal Anterior Cerebral Artery Aneurysm

*Gareth Rutter, Cristian Gragnaniello, Remi Nader, Anthony J. Caputy, Dimitri Sigounas, and Marguerite Harding*

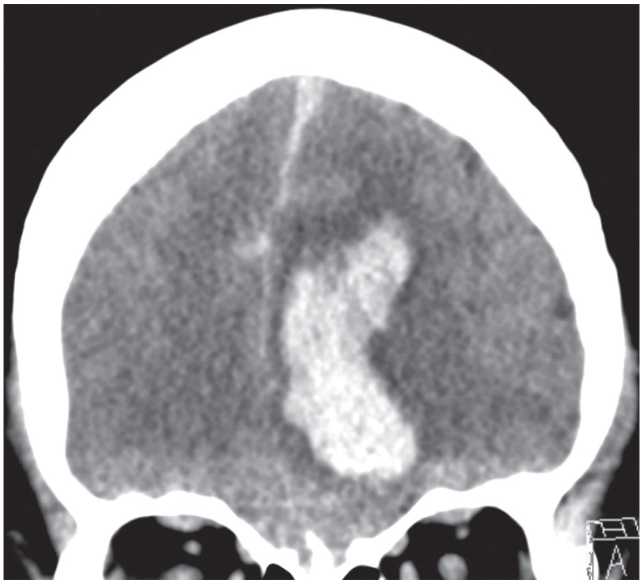

**Fig. 39.1** Coronal noncontrast CT.

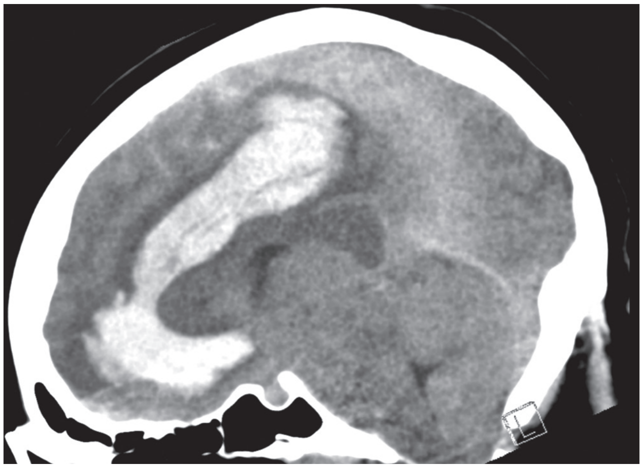

**Fig. 39.2** Sagittal noncontrast CT.

## Clinical Presentation

- A 46-year-old woman presents to the emergency room with sudden-onset headache and vomiting and reduced level of consciousness.
- On examination, she is confused and drowsy. She opens her eyes to voice and obeys commands. There is a dense weakness of her right lower limb.
- A CT scan is performed in the emergency room (▶**Fig. 39.1**, ▶**Fig. 39.2**, and ▶**Fig. 39.3**).

## Questions

1. Describe the CT scan, including the Fisher grade.
2. What is the diagnosis? What is the World Federation of Neurosurgical Societies (WFNS) grade?
3. How will you work up this patient? What further investigations should be considered?
4. Describe the anatomy of the anterior cerebral artery (ACA). What is the *relative* incidence of distal ACA (DACA) aneurysms at each division?
5. What are the typical morphological characteristics of DACA aneurysms and associated vascular abnormalities?
6. What is the role for endovascular management?
7. Describe potential surgical approaches and their limitations.

## Answers

1. *Describe the CT scan, including the Fisher grade.*
   - There is diffuse subarachnoid hemorrhage in the interhemispheric and pericallosal cisterns, and intracerebral hematoma within the left frontal lobe. The incidence of intracerebral hematoma is higher in ruptured DACA aneurysms (40–70% of cases) than aneurysms at other locations.[1]
   - The patient has a Fisher grade 4.[2]
2. *What is the diagnosis? What is the World Federation of Neurosurgical Societies (WFNS) grade?*
   - Ruptured aneurysm of the DACA
   - The patient has a Glasgow Coma Scale of 13 (E3V4M6) and has WFNS grade 3.[3]
3. *How will you work up this patient? What further investigations should be considered?*
   - Work-up includes admission to the intensive care unit, hemodynamic monitoring with arterial and central venous access, vasospasm prophylaxis, consideration of cerebrospinal fluid (CSF) diversion (balanced against the risk of aneurysm rerupture), and early definitive intervention (within 48 hours of admission). See Case 35, Carotid Cavernous Sinus Fistulas, for further details.
   - Digital subtraction angiography should be strongly considered.
4. *Describe the anatomy of the anterior cerebral artery (ACA). What is the relative incidence of distal ACA (DACA) aneurysms at each division?*
   - The ACA has been variously divided into three to five divisions. Microsurgically, it can be divided into following five anatomic divisions:[4,5]
     - A1 (precommunicating): bifurcation of internal carotid artery to anterior communicating artery (ACOM)
     - A2 (infracallosal): ACOM to genu of corpus callosum
     - A3 (precallosal): genu of corpus callosum to horizontal portion of ACA
     - A4 (supracallosal): horizontal portion of ACA to a plane through the coronal suture
     - A5 (postcallosal): a plane through the coronal suture to terminal cortical branches
   - Distal ACA aneurysms are classified into three divisions:
     - Proximal pericallosal aneurysms (aneurysms of A2): location of 6% of DACA aneurysms
     - Classic pericallosal aneurysms (aneurysms of A3): location of 85% of DACA aneurysms, predominately at the pericallosal–callosomarginal artery junction
     - Distal pericallosal aneurysms (aneurysms of A4, A5, and branches of A3, including callosomarginal artery): least common location of DACA aneurysms
5. *What are the typical morphological characteristics of DACA aneurysms and associated vascular abnormalities?*
   - DACA aneurysms tend to be smaller than average at rupture with a mean size of 5 to 8 mm. Therefore, elective intervention (surgical or endovascular) may be justified for unruptured DACA aneurysms < 7 mm.
   - DACA aneurysms usually have a broad base and often have a sclerotic or calcified neck.[6] These morphological characteristics pose technical limitations on both endovascular and microsurgical management options.[7]
   - Associated ACA vascular abnormalities occur in up to 35% of cases. The most common is an azygos ACA—up to 22% of cases. A concomitant arteriovenous malformation is associated in up to 15% of cases.[5] Multiple aneurysms occur in 20% of cases.
6. *What is the role for endovascular management?*
   - The International Subarachnoid Aneurysm Trial was not powered for DACA aneurysms; there are few studies comparing clinical outcomes of endovascular to microsurgical management in the DACA aneurysm population. One series found a statistically nonsignificant difference in clinical outcomes favoring microsurgery.[8]
   - Complete endovascular occlusion has been reported in 50% of cases, and near complete in 45%.[9] Recurrence has been documented at a rate of 18 to 22%.
   - Morphological characteristics increase the technical difficulty of coiling, with reported rates of periprocedural aneurysm rupture (12%) higher than for aneurysms at other sites.[10]
7. *Describe potential surgical approaches and their limitations.*
   - Each division of the DACA requires modification to the surgical approach. In general, the more proximal the DACA aneurysm, the more anterior the approach. Several morphometric anatomical landmarks have been described to assist in selection of the approach; the most commonly described being the relationship of the aneurysm to the genu of the corpus callosum.
   - Compared to aneurysms at other locations, DACA aneurysms have a higher than average incidence of intraoperative rupture. This is due to adherence of the aneurysm dome to the parenchyma and projection of the dome toward the surgeon, which predisposes them to rupture with excessive lobe retraction. Intraoperative rupture rates have been reported as high as 50%.
   - Infracallosal aneurysms:
     - Unilateral anterior frontal craniotomy, anterior interhemispheric approach
       - Most commonly published approach for infracallosal aneurysms
       - Modification of the approach using a morphometric landmark to determine optimal position of craniotomy, based upon relationship of the aneurysm to the long axis of the inferior segment of A2, has been described.[11]

## Answers (*continued*)

    - Difficult to establish proximal vascular control, which is exacerbated by the aneurysmal dome often being the first aspect of the aneurysm encountered. An anterior callosotomy may be considered to facilitate access to proximal ACA trunk.[5,11]
  - Lateral supraorbital approach
    - Suitable if aneurysm < 15 mm from anterior cranial base in vertical dimension
    - May require partial resection of gyrus rectus to visualize the aneurysm
- Supracallosal aneurysms:
  - Parasagittal frontal craniotomy, anterior interhemispheric approach
    - Narrow and deep surgical corridor within interhemispheric cistern, with an absence of anatomical landmarks—exacerbated by subarachnoid hematoma. Neuronavigation may mitigate absence of anatomical landmarks.
    - Limited CSF drainage from pericallosal cistern—may require adjunct ventriculostomy or lumbar puncture
    - Bridging veins can obstruct the surgical corridor, which may require sacrifice. Preoperative venogram may be considered to evaluate venous collateralization and the risk of postoperative venous infarction.[12]

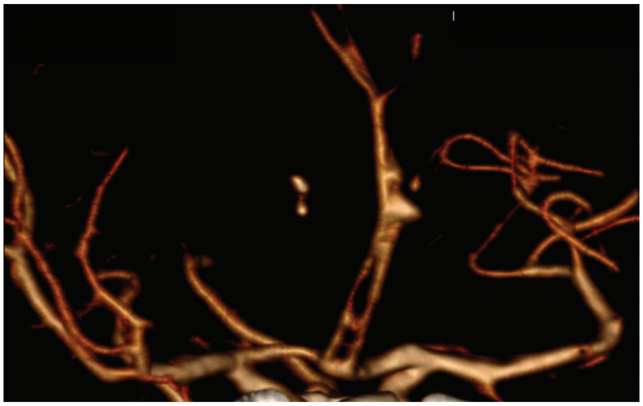

**Fig. 39.3** CT angiogram 3D reconstruction.

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# Case 40 Blister Carotid Aneurysm

*Nancy McLaughlin and Michel W. Bojanowski*

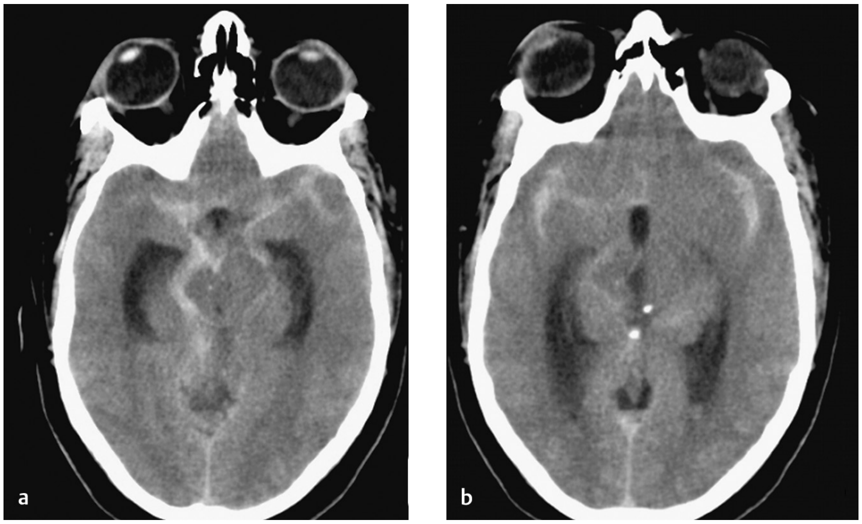

**Fig. 40.1** (**a, b**) CT scan of the head showing a subarachnoid hemorrhage.

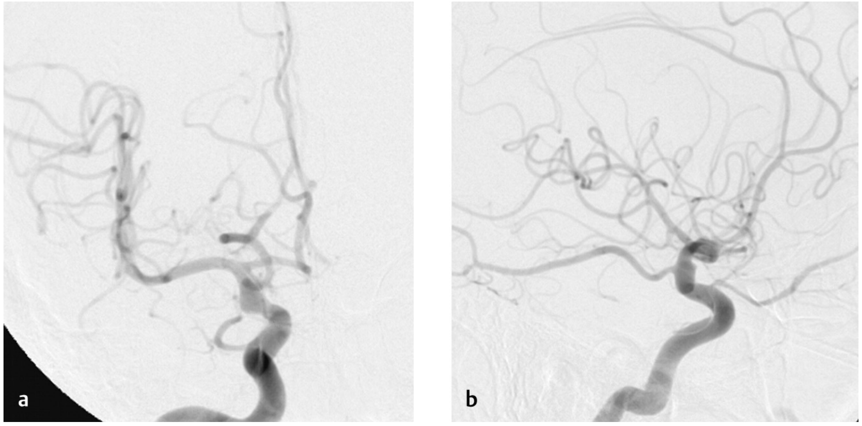

**Fig. 40.2** Cerebral angiography. (**a**) Anteroposterior and (**b**) lateral views of a right internal carotid.

## Clinical Presentation

- A 42-year-old woman presents with a sudden-onset headache. Her past medical history is unremarkable and she is not on any regular medication.
- Her vital signs are stable and the physical examination is normal. Blood pressure is 125/80 mm Hg.
- Except for drowsiness, her neurological examination is unremarkable.
- A CT scan of the head is performed as shown in ▶**Fig. 40.1**. Subsequently, a four-vessel cerebral angiography (▶**Fig. 40.2**) reveals an ectatic appearance of the supraclinoidal segment of the right internal carotid artery (ICA) with possibly a faint bulge at the level of the posterior communicating artery. The angiogram is otherwise normal.

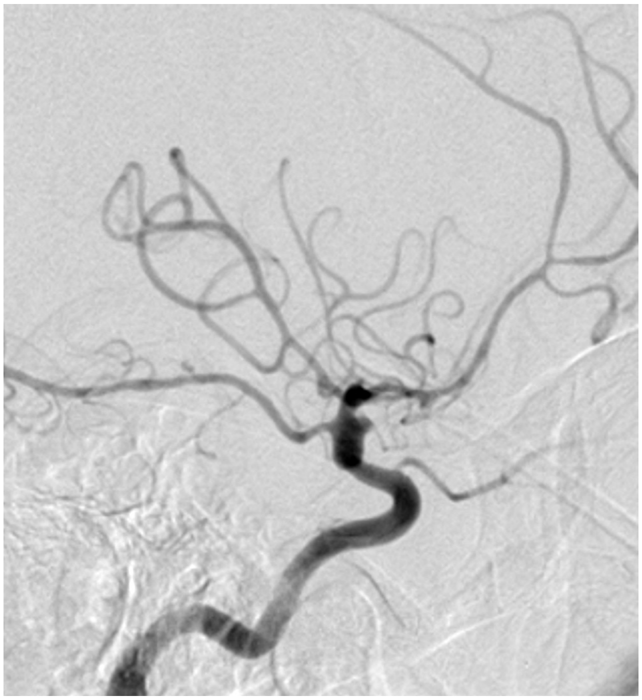

**Fig. 40.3** Cerebral angiography: lateral view, right internal carotid done 3 days later.

## Questions

1. What are the possible causes of subarachnoid hemorrhage (SAH) with a negative angiogram?
2. What are the CT criteria for pretruncal nonaneurysmal SAH?
3. What is your initial management?
   You decide to repeat the angiogram 3 days later (▶**Fig. 40.3**).
4. Interpret the angiogram.
5. What is the definition of this lesion and how can it be categorized?
6. What are the pathological features?
   After discussion with the neurointerventional team, you recommend surgical treatment.
7. What surgical strategies should be taken into consideration?
8. How do blister aneurysms differ from very small (< 3 mm) saccular aneurysms?
9. How does the treatment of very small aneurysms differ from that of blister aneurysms?
10. What is the prognosis of blister aneurysms?

## Answers

1. *What are the possible causes of subarachnoid hemorrhage (SAH) with a negative angiogram?*
   - Aneurysm not seen on initial angiogram in following cases:
     - Incomplete or suboptimal quality images
     - Very small aneurysms
     - Thrombosis of an aneurysm after SAH
     - Lack of filling due to vasospasm
     - Nonaneurysmal SAH
   - Pretruncal nonaneurysmal SAH
     - Angiographically occult vascular malformations including cavernous malformations
     - Coagulation disorders
     - Drug abuse (e.g., cocaine)
     - Cerebral artery dissection (e.g., vertebral intracranial)
     - Pituitary apoplexy
2. *What are the CT criteria for pretruncal nonaneurysmal SAH?*
   - Epicenter of hemorrhage anterior to the brainstem (interpeduncular and/or prepontine)
   - There may be extension into the anterior part of the ambient cistern or the basal part of the Sylvian fissure.
   - Absence of complete filling of the anterior interhemispheric fissure
   - No more than minute amounts of blood in the lateral portion of the Sylvian fissure
   - Absence of frank intraventricular hemorrhage: small amounts of blood sedimenting in the occipital horns of the lateral ventricle is permissible.[1]

## Answers (*continued*)

3. *What is your initial management?*
   - Please refer to Case 44 [Stent and balloon assisted coiling] and Case 36 [SAH and vasospasm] for a more detailed approach to this answer.
   - The CT scan has shown an aneurysmal SAH distribution and does not satisfy the criteria of pretruncal SAH mainly because of the filling of the Sylvian and interhemispheric fissures. Accordingly, a ruptured intracranial aneurysm is not entirely excluded. Note the presence of moderate hydrocephalus. The initial management includes:
     - Admission to intensive care unit
     - External ventricular drain (EVD) if there is progression of hydrocephalus or clinical deterioration
     - Bed rest and symptomatic treatment (e.g., analgesic, antiemetic)
     - Control of blood pressure
     - Cardiac monitoring
     - Intravenous (IV) fluids: normovolemia
     - Calcium channel blockers: nimodipine
     - Lab work: arterial blood gas (ABG), electrolytes, complete blood count (CBC), international normalized ratio (INR), partial thromboplastin time (PTT)
4. *Interpret the angiogram.*
   - The right ICA injection reveals a small bulbous, broad-based dilatation at the anterior wall of the right ICA.
   - This finding is compatible with the diagnosis of a ruptured blister ICA aneurysm. These aneurysms reportedly exhibit rapid growth and changes in shape, as in this case.[2]
5. *What is the definition of this lesion and how can it be categorized?*
   - Blister-like aneurysms are often small sessile, hemispherical expansions at a nonbranching site of the anterior aspect of supraclinoid segment of the ICA. However, they are known to be heterogeneous, presenting a broad spectrum of morphology, and may, at times, be large.[3]
   - They are characterized by a very fragile wall and a poorly defined neck and are histologically distinct from saccular aneurysms.
   - In relation to the ICA, they have been referred to as:
     - Dorsal
     - Distal-medial
     - Superior
     - Anterior wall
6. *What are the pathological features?*
   - These aneurysms are sometimes associated with arteriosclerosis of the neighboring carotid wall.
   - Abrupt termination of the internal elastic lamina is seen at the border between the normal and sclerotic carotid wall.
   - The dome is composed of fibrinous tissue and adventitia, while the usual collagenous layer of saccular aneurysm is absent.[4]
   - They occur probably secondary to a subadventitial dissection of the ICA.
   - Due to their very fragile walls and poorly defined necks, surgical exploration and clipping are very hazardous with a high rate of intraoperative or postoperative rupture.[5]
7. *What surgical strategies should be taken into consideration?*
   - Blister aneurysms have a high risk of intraoperative rupture with lacerations of the ICA during clipping. These aneurysms require different surgical techniques and strategies based on their heterogeneous morphology.[3]
   - Strategies include:[2,3,5,6]
     - Exposure of the cervical ICA for proximal control if deemed necessary
     - Opening of the Sylvian fissure before accessing the carotid and chiasmatic cisterns
     - Particular care to minimize frontal lobe retraction because of possible adhesions of the frontal lobe to the aneurysm dome
     - Clipping while pressure within the ICA is low with temporary clipping of the ICA
     - Clip blades should be applied parallel to the parent artery and should include part of the healthy wall.
     - Confirming the intraoperative stability of clips is essential and is done with induced blood pressure elevation and repeated irrigation before closing the dura mater.
   - Other potential surgical treatments
     - Wrapping the full circumference of the ICA and applying a clip (clipping over wrapping)
     - ICA trapping with or without bypass
     - Direct suturing
     - Endovascular strategies:
       - ○ Coiling: hazardous despite the use of softer coils and balloon-assisted techniques, given the pathological features of blister aneurysms. It has not provided satisfactory results.[7]
       - ○ Stent-assisted coiling: it often requires a second treatment.[8] Stent-within-stent[9] and flow diverters[10,11] are being used more frequently. However, they require dual antiplatelet therapy, which can be an issue in patients necessitating surgical procedures. Long-term results are not available at present.
8. *How do blister aneurysms differ from very small (< 3 mm) saccular aneurysms?*
   - It is critical to distinguish between these two types of aneurysms.[3,12]
   - Blister aneurysms are rare, whereas very small saccular aneurysms are a more frequent cause of SAH.
   - Unlike saccular aneurysms, blister aneurysms arise at a nonbranching site of the parent artery, most often in the anterior wall of the ICA.

## Answers (*continued*)

- Blister aneurysms are thought to result from a dissection of the main vessel, thus making them histologically distinct from saccular aneurysms. In blister aneurysms, the wall of the parent artery is also affected.
- Although blister aneurysms are often small, they are known to potentially increase rapidly in size and may at times be large.
- The nonrecognition in the difference of the diseased wall between the two aneurysms may be the reason for the reportedly high rate of morbidity and mortality related to treatment of blister aneurysms, whether by surgery or endovascular means.

9. *How does the treatment of very small aneurysms differ from that of blister aneurysms?*
   - The size of an aneurysm is a key factor in surgery; small saccular aneurysms being usually simpler to treat surgically than larger ones.
   - Contrary to blister aneurysms, saccular ones can be clipped at the neck, where sufficient tissue remains to hold the clip, without the need to include part of the healthy wall of the parent artery.
   - However, intraoperative rupture has been more commonly reported for very small aneurysms compared to larger ones, which may affect the outcome.
   - When using endovascular procedures, very small aneurysms are reported to have a higher risk of rupture and a higher mortality rate compared to larger ones.[13,14]
   - Recent reports suggest that these risks have been diminished with the use of flexible coils and balloon-assisted technique.[14,15,16]
   - Endovascular treatment of blister aneurysms carries a higher procedural risk compared to treatment of very small saccular aneurysms.
10. *What is the prognosis of blister aneurysms?*
    - The prognosis is related to the clinical status on admission, evaluated by the Hunt and Hess clinical classification.
    - Because of the high incidence of intraoperative or postoperative bleeding, the prognosis is markedly worse than for those patients with saccular type aneurysm.[5]

    However, accurate preoperative diagnosis of a blister aneurysm and adapting the strategy according to its morphology may result in excellent outcome.[3,6]

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# Case 41 Basilar Tip Aneurysms

*Anthony M. T. Chau, Peter J. Mews, Aaron S. Gaekwad, Marguerite Harding, and Cristian Gragnaniello*

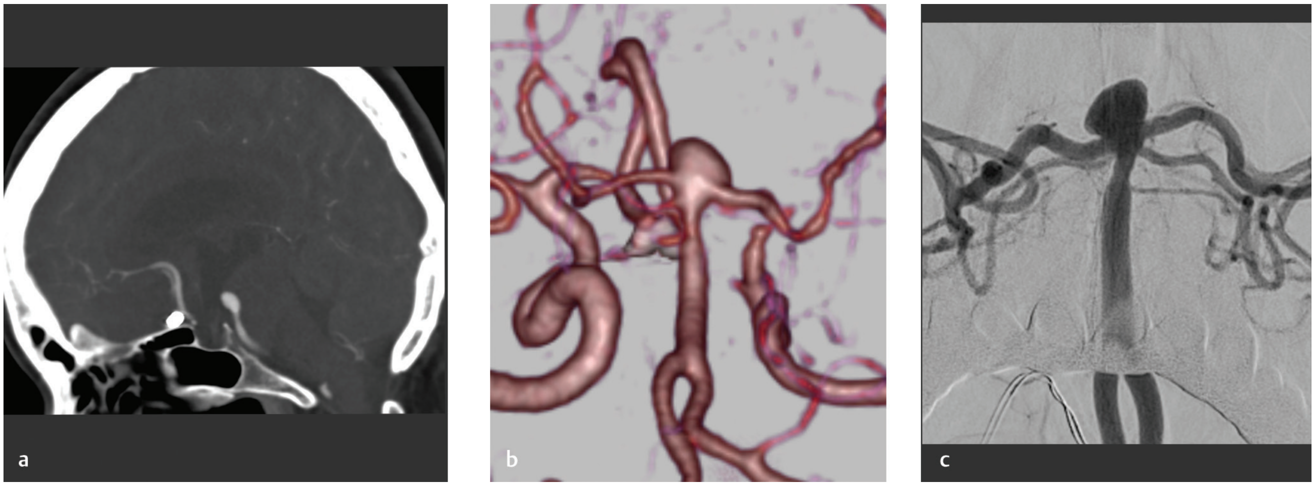

**Fig. 41.1** (**a**) Sagittal CT angiogram; (**b**) coronal 3D reconstruction CT angiogram; (**c**) coronal digitally subtracted angiogram.

## Clinical Presentation

- A 63-year-old Caucasian woman follows up in your clinic for management of an unruptured 7-mm basilar tip aneurysm (BTA; ▶**Fig. 41.1**).
- It was discovered 1 year prior when she suffered a WFNS grade 2 subarachnoid hemorrhage (SAH) secondary to a ruptured 6-mm anterior communicating artery (ACommA) aneurysm. This was treated with an external ventriculostomy followed by endovascular coiling.
- She presents for your review having made a complete neurologic recovery, with follow-up imaging having demonstrated no recurrence of the ACommA aneurysm and no growth in the BTA.
- She is a nonsmoker with a history of controlled hypertension.

## Questions

1. Describe the radiological features.
2. What is the epidemiology of BTAs?
3. What is the annual risk of rupture of unruptured BTAs?
4. Describe the anatomy of the basilar apex.
5. What anatomical features are thought to contribute to BTA formation and rupture?
6. Outline the broad considerations for determining microsurgical versus endovascular intervention.
7. Outline specific surgical considerations.
8. What is your management of an intraoperative perforator injury?
9. What functional outcomes may be expected?

## Answers

1. *Describe the radiological features.*
   - There is an anterosuperiorly projecting saccular aneurysm arising from the basilar apex, measuring 7 mm in maximal diameter. It sits in the interpeduncular cistern above the level of the posterior clinoid process.
   - It has a relatively wide neck measuring 4 mm, giving a dome-to-neck ratio of 1.75. The height of the aneurysm is 6 mm giving an aspect ratio of 1.5. The aneurysm wall is smoothly contoured with no daughter sacs.
   - The posterior cerebral arteries (PCAs) arise at the base of the aneurysm, at an angle perpendicular to the feeding basilar artery. The basilar artery harbors a mild stenotic segment just proximal to the superior cerebellar arteries.
2. *What is the epidemiology of BTAs?*
   - Three percent of the population harbors unruptured intracranial aneurysms (UIAs); BTAs account for 5 to 7% of these and 50% of the posterior circulation aneurysms.[1–3]
   - Typically detected in the fifth to sixth decades of life; more common in women. Presentation may be incidental, or with headaches, symptoms from mass effect, or SAH.
   - BTAs are particularly prone to rupture compared with anterior circulation aneurysms (13.8 annual relative risk of rupture).[4]
3. *What is the annual risk of rupture of unruptured BTAs?*
   - Specific data from Japanese cohort of newly diagnosed, unruptured BTA aneurysms (*n* = 445 BTAs, largest published in the literature):[2]
     - 3 to 4 mm: 0.23%
     - 5 to 6 mm: 0.46%
     - 7 to 9 mm: 0.97%
     - 10 to 24 mm: 6.94%
     - ≥25 mm: 17.82%
   - The PHASES score for unruptured aneurysms takes into account posterior circulation location, ethnicity (North American/European vs. Finnish vs. Japanese), size of aneurysm, hypertension, and history of SAH in a 5-year risk prediction chart.[5]
     - This Caucasian (non-Finnish) patient has a history of ACommA-related SAH, hypertension, and a 7-mm unruptured BTA. Her PHASES score demonstrates intermediate risk for rupture, specifically a 6% risk for rupture within the next 5 years.
4. *Describe the anatomy of the basilar apex.*
   Anatomy of the basilar apex, major branches, and perforators[6–8] are as follows:
   - BTAs can arise from the basilar apex, proximal PCAs, or proximal superior cerebellar arteries (SCAs).
   - The basilar apex sits in the interpeduncular and upper part of the prepontine cisterns.
     - Its location in relation to the posterior clinoid process varies. In half of cases, it sits at the level of the dorsum sellae and posterior clinoid process. Otherwise, it sits above or below this level.
     - As the PCA must pass above the oculomotor nerve and tentorium cerebelli, the vertical location of the basilar apex affects the angulation of the PCA and the proximity of the perforators to the aneurysm dome.
   - The SCAs are the first branches located caudal to the PCAs, the terminal bifurcation of the basilar apex. The SCA courses below the oculomotor nerve and is duplicated in 10% of cases. Seven variations in the origin of the SCA have been described by Yasargil.[9]
     - A: both arise as a single vessel separate from the PCAs
     - B: both arise from the bifurcation of the basilar artery (BA) into the PCAs
     - C: each SCA arises from the PCAs
     - D: one SCA arises from the PCA, the other from the BA
     - E: one SCA arises from the bifurcation of the BA, the other from the PCA
     - F: duplicate SCAs arise from the BA bilaterally
     - G: duplicate SCAs arise from one side of the BA, and a single SCA arises from the contralateral side
   - The rostral BA, proximal PCAs, and proximal SCAs all harbor eloquent perforating vessels. These are end arteries, injury to these will induce potentially devastating infarcts.
     - Proximal PCA (P1): thalamoperforating group, consisting of mesencephalic and diencephalic vessels. Range from two to five in number but with numerous side branches, they extend through the interpeduncular cistern to enter the posterior perforated substance or median sulcus of the cerebral peduncle to supply the central and paramedian midbrain, and the medial part of the subthalamus and thalamus.
     - Rostral BA: interpeduncular, superior medial pontine, and lateral pontine groups. Usually arise from its lateral and dorsal surface, approximately three per side rostral to the SCA.
       - ○ The BA below the SCA origin usually harbors a perforator-free zone.
     - Proximal SCA: interpeduncular, superior/inferior medial and lateral pontine, basal cerebellar, lemniscal trigone groups. Typically, two to three perforators are present within the first 15 mm of the proximal SCA, which may be direct or circumflex in their course to the brainstem.
     - Anastomoses between the PCA, BA, and SCA branches (if they occur) are typically unilateral.

## Answers (*continued*)

  - The posterior communicating artery (PComA) most often only possesses one perforating branch, the premamillary artery, which enters the diencephalon between the cerebral peduncle, optic tract, and mamillary body. It can arise from the anterior, middle, or posterior part of the PComA, but rarely from the PComA–PCA junction.

5. *What anatomical features are thought to contribute to BTA formation and rupture?*
   - Geometric anatomic relationships associated with BTA formation[10,11]
     - Small basilar artery diameter (higher jet flow at the apex/bifurcation)
     - Large P1–P1 angle (the angle formed between the origins of the PCA arteries; greater divergence of flow and reduced wall shear stress—seems to increase the risk for rupture)
   - Geometric relationships associated with BTA rupture[2,11]
     - Large P1–P1 angle
     - Irregularity/presence of a daughter sac
6. *Outline the broad considerations for determining microsurgical versus endovascular intervention.*
   Considerations for determining microsurgical versus endovascular intervention are as follows:
   - Microsurgical:[12–14]
     - Microsurgical intervention traditionally achieves higher complete occlusion rates > 90%. It is more durable, has minimal recurrence, and is used particularly for younger patients.
     - Advantageous particularly for complex aneurysms: those with a wide neck > 4 mm, smaller dome-to-neck ratio, and a smaller aspect (height: neck) ratio
     - Preferred for large/giant aneurysms, especially those causing mass effect.
     - Less expensive, widely accessible in developing countries
     - However, it is technically very challenging; BTAs are historically associated with higher surgical morbidity compared with other operated aneurysms.[1]
   - Endovascular:[15–17]
     - It is a safer but potentially less efficacious or durable form of treating BTAs. Close serial monitoring and retreatment is often required.
     - Options include coiling alone (suboptimal results) or addition of parent artery reconstruction particularly for wide-neck aneurysms. Following techniques are used:
       - ○ Balloon remodeling
       - ○ P1–P1 horizontal stent-assisted via the PComA
       - ○ Single stent-assistance running from the BA to the PCA with greater aneurysmal neck involvement
       - ○ Y-stent-assisted coiling
     - Newer devices under development, including endosaccular flow-disrupting devices (Woven Endobridge [WEB] device, Sequent Medical, Aliso Viejo, CA; Medina, Medtronic neurovascular), also show promise for treatment of basilar apex aneurysms.
     - Stent diversion encourages stasis and thrombosis at the end-vessel basilar apex aneurysm. Ideally there will be endothelial regeneration over the neck of the aneurysm but this may be difficult to achieve.[18]
       - ○ Y-stents are particularly useful in the treatment of wide-neck BTAs.
       - ○ Current evidence suggest lower recanalization, retreatment, and rebleed rates in BTAs managed with stent-assisted coiling versus coiling alone.
       - ○ Major concern of stent use is thrombogenicity/infarct, so perioperative dual antiplatelet therapy with close monitoring of platelet function inhibition is generally required.
     - Ruptured or unruptured aneurysm
       - ○ Ruptured: traditionally a detractor is used for stenting due to need for dual antiplatelets, unless in the rescue situation.
       - ○ Some endovascular centers will administer loading doses at the time of intervention for ruptured BTAs. In one endovascular series, thromboembolism developed in one out of three ruptured BTAs treated acutely with a Y-stent.[17]
     - Improving but variable data with newer technology
       - ○ One contemporary endovascular series of BTAs treated with coil and stent-assisted coiling achieved complete or “near complete” occlusion rates of 88%.[15]
       - ○ A recent literature review of BTAs treated with Y-stents reported complete occlusion rates of 0 to 95% (most series reported results in the 20–50% range)[17]

   Complications:
   - Microsurgical:[13,14]
     - Procedure-related deaths: 3 to 4%
     - Ophthalmoplegia, particularly oculomotor nerve: 46% transient, 6% permanent
     - Perforator injury: 19%
     - P1 stenoses: 11%, may occur particularly if the basilar apex is high and the proximal PCAs slope downward
     - Further intervention: 5%
   - Endovascular:[15,17,19]
     - Procedural-related deaths: 0 to 2%
     - Thromboembolism: 7 to 12%
     - At 2-year follow-up

## Answers (*continued*)

    - Recurrence/recanalization: 30% overall; 17% stent-assisted, 39% coil only
    - Rebleed: 4% stent-assisted, 3 to 5% coil only
    - Further intervention: 20% overall; 8% stent-assisted, 28% coil only

7. *Outline specific surgical considerations.*
- Patient age, comorbidities, and performance status
- Experience of the surgical team
- Aneurysm morphology:
  - Size
    - Occlusion rates after coiling of large/giant BTAs: 40%.[19] Endovascular options do not address mass effect.
  - Neck width, dome-to-neck ratio, aspect ratio
    - Wide-neck aneurysms > 4 mm: microsurgical clipping is usually preferred; alternatives include stent-assisted coiling with pipeline or Y-stents
    - Occlusion rates after coiling of wide-neck BTAs: 40%.[19] But with the advent of flow-diverting stents, wide-neck and large/giant BTAs may prove to be adequately addressed, with occlusion rates > 90%.[20]
  - Major vessels coming off the aneurysm neck
  - Wall irregularity, calcification/atherosclerosis
  - Presence of aneurysmal thrombus
  - Angulation/direction of dome:[8]
    - Anteriorly projecting: points toward the posterior clinoid process, manipulation and visualization of the basilar trunk for proximal control may be challenging.
    - Superiorly projecting: favored projection for surgical clipping, particularly if it is not too highly located
    - Posteriorly projecting: least favored for surgical clipping, as the perforators of the P1 segment may be draped over the anterior aspect of the aneurysm, and the proximal SCA perforators around its posterior aspect
  - Vertical distance of aneurysm above or below the posterior clinoid process; retro- or subsellar location: influences the surgical approach taken (see below).
- Presence of nonfetal-type circulation of PComA: this would allow the sacrifice of the PComA for increased intraoperative exposure.
- Presence and location of other aneurysms: whether these may be treated at the same sitting.

Surgical approaches:[8,12,14,21]

- Frontotemporal/transsylvian (▶**Fig. 41.2**)
  - Favored for high BTAs, wide P1–P1 angle, aneurysmal calcification, or thrombus
  - Orbitozygomatic craniotomy, wide Sylvian fissure split, resection of posterior clinoid process, and occasionally opening of the cavernous sinus to increase visualization of the interpeduncular fossa
  - Long, narrow working corridor
  - Sectioning of nonfetal-type/hypoplastic PComA near the PCA junction (perforator-free zone) may be performed to increase exposure, mobilize ICA, and enhance visualization of the P1 perforators. Only performed if P1 is larger than the PComA.
- Subtemporal
  - Favored for retrosellar BTAs and those with a narrow P1–P1 angle.
  - Perpendicular view of the aneurysm neck including its posterior aspect is afforded. Brainstem perforators, PCAs, and SCAs are well visualized.
  - Short working distance but contralateral P1 may be difficult to visualize.
  - Tentorial incision for sufficient exposure, although there is risk to trochlear nerve.
  - Requirement for retraction may be problematic in the ruptured setting and may result in brain contusions or injury to the vein of Labbé; can be managed with gentle retraction, cerebrospinal fluid (CSF) release/lumbar drainage, and occasionally temporal partial lobe Tony.
- Presigmoid
  - More invasive approach, favored for subsellar BTAs
  - Combined middle fossa/infratentorial approach, drill down of mastoid and petrous bone with preservation of semicircular canals, presigmoid and middle fossa dural opening, division of petrous sinus, and opening of tentorium behind the insertion of the trochlear nerve.

8. *What is your management of an intraoperative perforator injury?*
- Injury to brainstem perforators should be avoided by meticulous dissection techniques. The posterior wall of the aneurysm should be fully visualized before positioning of the final clip.
- Manipulation of the perforators should be kept to a minimum and use of cautery should be avoided as their injury is oftentimes unforgiving.
- When applying the permanent clip, if a perforator is included in this clipping, then it should be repositioned and papaverine be applied to the area of the perforators prior to closure.
- If infarction in the territory of the perforator persists postoperatively, then following standard measures for postoperative stroke treatment should be initiated:
  - Obtain CT and angiogram: consider papaverine or verapamil intravascular injection
  - Stabilize in intensive care unit (ICU):
    - Airway and ventilator support,
    - Hemodynamic monitoring and IV hydration.
    - Central line and arterial line placement.
    - Supplemental oxygen.

## Answers (*continued*)

    - Neuro checks
    - Keep supine with modest head elevation
  - Maintenance of:
    - Normothermia
    - Normoglycemia
    - Normotension
  - Laboratory studies: complete blood count (CBC), electrolytes, coagulation profile
  - Correct anemia if present: consider blood transfusion if necessary
  - Consider antiplatelet therapy > 24 hours after craniotomy

9. *What functional outcomes may be expected?*
   Good functional outcomes at 1 year (Modified Rankin Score 0–2):[13]
   - Ruptured
     - Microsurgical: 73%
     - Endovascular: 77%
   - Unruptured
     - Microsurgical: 92%
     - Endovascular: 91%

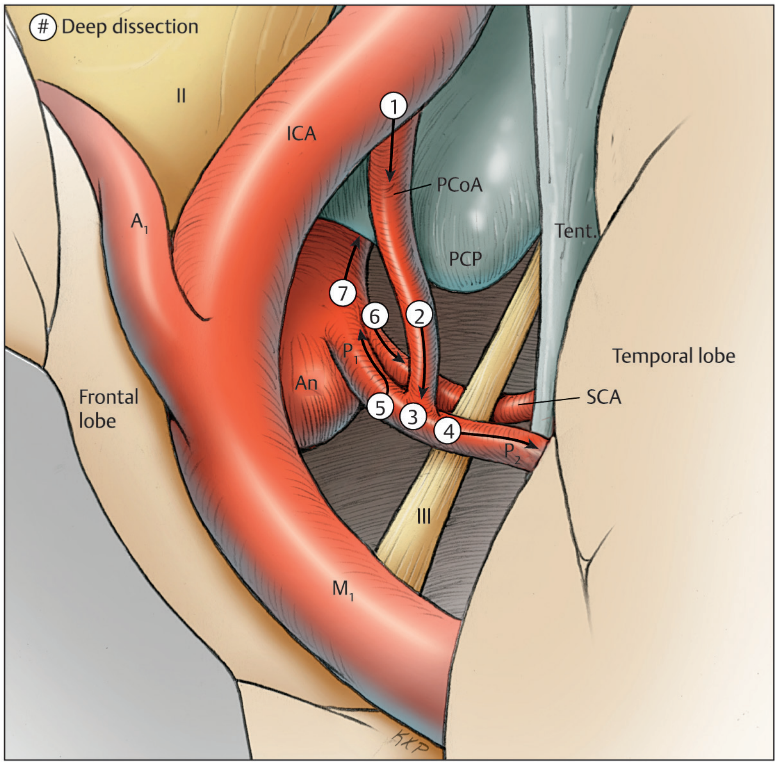

**Fig. 41.2** Pterional exposure of basilar apex aneurysm. (Reproduced with permission from Lawton M. Basilar artery bifurcation aneurysms. In: Lawton M, ed. Seven Aneurysms. 1st ed. New York: Thieme; 2011:169. Figure 19.5)

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# Case 42 Vertebrobasilar Junction Aneurysms

*Isabella Esposito, Cristian Gragnaniello, and Marguerite Harding*

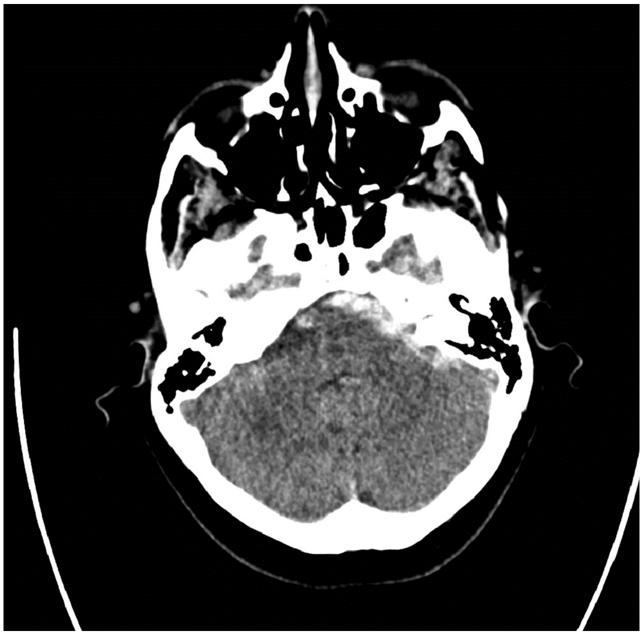

**Fig. 42.1** Preoperative imaging. Axial CT scan.

## Clinical Presentation

- A 54-year-old man presents to the emergency department (ED) with sudden onset of occipital headache.
- An emergent CT scan of his brain which prompted a CT angiogram demonstrated a ruptured left vertebrobasilar junction (VBJ) aneurysm (▶**Fig. 42.1** and ▶**Fig. 42.2**).

## Questions

1. Describe the imaging.
2. What symptoms are associated with VBJ aneurysms?
3. What would the rest of the radiographic work-up include?
4. Briefly describe the vascular anatomy of the VBJ, including variations and perforators where relevant.
5. Briefly describe the embryological basis for basilar artery fenestration.
6. What are the management options of this condition?
7. What surgical options can you offer to a patient with a VBJ aneurysm not amenable to an endovascular treatment? What are the complications and outcomes?
8. Describe the endovascular treatment options of VBJ aneurysms.
9. Describe the role of anastomotic revascularization procedures in the management of VBJ aneurysms.
10. You elect a surgical repair via clipping. The procedure is uneventful; however, the patient wakes up with hoarseness and difficulty in swallowing. How do you explain and proceed with this occurrence?

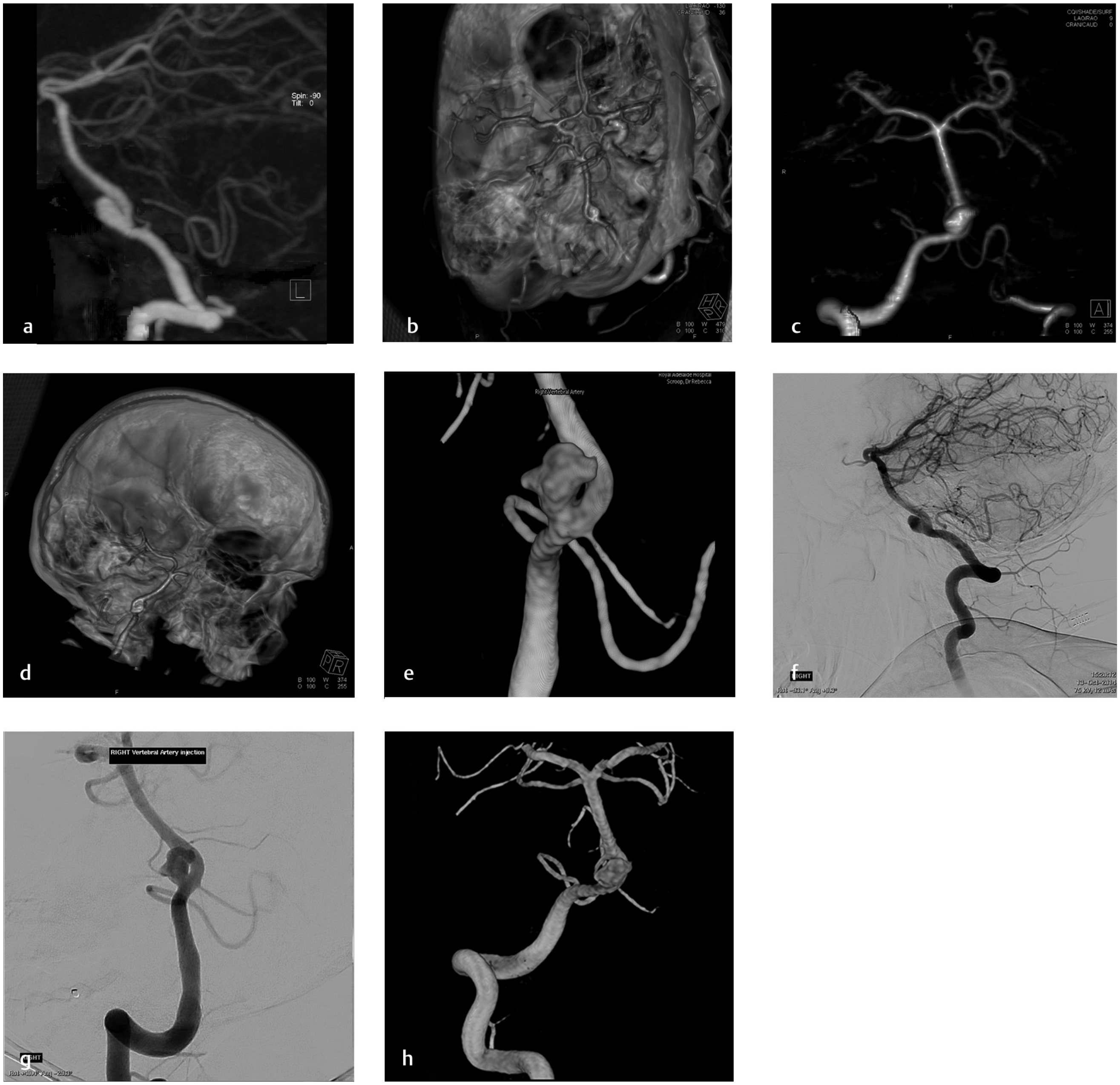

**Fig. 42.2** (**a–h**) High-resolution CT with three-dimensional reconstructions and digital subtraction angiography highlight a ruptured, large saccular lobulated aneurysm, originating from the vertebrobasilar junction. The basilar artery fills from the left vertebral artery.

## Answers

1. *Describe the imaging.*
   - Axial CT scan obtained on admission reveals a subarachnoid hemorrhage (SAH) with blood in the basal cisterns not extending to the fourth ventricle.
   - At this stage, prior to further vascular imaging and given the location of the blood, an aneurysm originating either from the basilar artery or the VBJ cannot be excluded. CT angiography and a conventional angiogram shows a ruptured, large saccular lobulated aneurysm, originating from the vertebrobasilar junction. The basilar artery fills from the left vertebral artery. The aneurysm is pointing in rostral and superior direction.
2. *What symptoms are associated with VBJ aneurysms?*
   - Most patients present with SAH and their condition can range in symptoms from mild headache to frank coma.
   - Infarction and, in rare cases, brainstem compression can also be part of the presenting picture and clinically range from subclinical dysfunctions to overt clinical signs. Following are the cardinal features to look for:
     - Ipsilateral peripheral cranial nerve involvement

## Answers (*continued*)

  - Contralateral weakness
  - Cerebellar signs, if present, are usually ipsilateral

3. *What would the rest of the radiographic work-up include?*

   Imaging investigations include:
   - CT scan and high-resolution CT with three-dimensional (3D) reconstruction
   - CT angiogram
   - Digital subtraction angiography (DSA) and rotational angiography with 3D rendering, if available

   Imaging should clarify the following features:
   - The blood distribution within the basal cisterns
   - The size, shape, and orientation of the aneurysm to differentiate between saccular, fusiform, and dissecting lesions as well as its exact location in relation to the VBJ and the midline
   - The relationship to the major surrounding skull base structures (main anatomical landmarks for orientation during surgery: the most lateral point of the foramen magnum, the dural entrance of the vertebral artery (VA), the posterior condylar canal, the distal sigmoid sinus, the hypoglossal canal and, in rare cases of high riding VBJ, the jugular tubercle) as well as the degree of involvement of the brainstem and rootlets of the lower cranial nerves
   - VA dominance and collateral branches: obtain a preoperative dynamic vascular study, such as a balloon test, occlusion test, or an Allcock test, to evaluate collateral flow from the opposite VA or the posterior communicating artery. This knowledge is critical in the instances of aneurysm trapping or prolonged temporary clipping.
   - The presence of a basilar artery fenestration (on 2D angiographic images, a small fenestration may be easily overlooked on other imaging modalities), which increases the risk of inadequate clip application
   - The optimal working projection for coiling (3D rotational angiography)
   - The presence of multiple intracranial aneurysms or an associated arteriovenous malformation

     In this case CT scan and subsequent DSA confirm the diagnosis of a ruptured, large saccular lobulated aneurysm originating from the VBJ. The basilar artery fills from the left VA.

4. *Briefly describe the vascular anatomy of the VBJ, including variations and perforators where relevant.*
   - The VBJ may be located a few millimeters above to 15 mm below the level of the pontomedullary junction.
   - The morphology of the main arteries is characteristically variable (i.e., duplications, fenestrations) whereas the perforators are usually consistent, particularly in terms of their numbers and points of penetration into the brain substance. Four groups of lower brainstem perforating arteries have been identified:
     - Group I: from the proximal VA to the posterior olivary sulcus
     - Group II: from the posterior inferior cerebellar artery (PICA) and distal VA to the posterior olivary sulcus
     - Group III: from the VBJ and the anterior inferior cerebellar artery to the superior olivary groove
     - Group IV: from the VBJ to the region of the foramen cecum
   - As mentioned, the VA may vary and may be hypoplastic, aplastic, duplicated, fenestrated, or it may make abnormal anastomosis with the internal carotid artery (ICA). VA may terminate in the PICA, and, in such cases, the basilar artery originates from the contralateral VA.
   - A hypoplastic VA can give rise to a dominant and singular anterior spinal artery (ASA). Unilateral origin of the ASA has been reported in about 10% of cases.

5. *Briefly describe the embryological basis for basilar artery fenestration.*
   - In the fetus, the paired longitudinal neural arteries that lie on either side of the developing hindbrain (aka rhombencephalon) slowly oppose each other and, during the fifth gestational week, fuse in the midline, between the level of the primitive trigeminal and hypoglossal arteries.
   - At the caudal end of the longitudinal neural arteries, temporary bridging arteries regress as fusion is completed and their persistence results in fenestration of the basilar artery.
   - At level of the fenestrated artery, the intrinsic architecture of its lateral walls is normal while the medial walls have focal defects of the tunica media that are frequently associated to aneurysm formation.

6. *What are the management options of this condition?*
   - The management options, as in all ruptured saccular aneurysms, include a conservative, endovascular and surgical approach.
   - The choice is made looking at both the characteristics of the aneurysm and the patient comorbidities.
   - These aneurysms are more commonly treated with endovascular strategies; however, in cases where endovascular therapies are not possible, available, or fail the patient should be referred to an experienced cerebrovascular neurosurgeon with the common goal of managing the obliteration of the aneurysm with minimal risk, both short and long term, to the patient.

7. *What surgical options can you offer to a patient with a VBJ aneurysm not amenable to an endovascular treatment? What are the complications and outcomes?*
   - A VBJ aneurysm can be exposed through a variety of approaches, depending on the characteristics of

## Answers (*continued*)

the lesion and individual surgical experience as for all lesions; most commonly a far lateral approach ipsilateral to the lesion is utilized.

- In a basic retrocondylar far lateral approach, positioning is of utmost importance as it allows for maximal opening of the posterior cervical suboccipital angle. This is achieved with two key modifications of the standard park bench (aka lateral recumbent) position:
  - The contralateral arm placed in a dependent fashion below the level of the body
  - The head positioned with the sagittal suture parallel to the floor and, then, flexed and rotated to place the inferior clivus perpendicular to the floor (Spetzler in Schmidek 6th ed).
- Condylar resection is tailored to the individual patient and appears to be helpful in the following cases: large condyles, an elliptical foramen magnum with a long anteroposterior diameter, and narrow-width or a lower lying clivus.
- Complications of approaches in this area include:
  - Transient or permanent lower cranial nerves dysfunction
  - VA injury (retraction related occlusion, laceration, pseudoaneurysm associated to temporary clipping) with sequelae varying from asymptomatic to severe (brainstem or cerebellar infarction, quadriparesis, death, delayed facial paralysis) depending on VA dominance and collateral branches.

8. *Describe the endovascular treatment options of VBJ aneurysms.*
   - The standard endovascular treatment is primary coil embolization with good initial and midterm results in small- and mid-sized VBJ aneurysm, whether related or not to a proximal basilar fenestration; in selected wide-neck aneurysms balloon remodeling (temporary inflation of a nondetachable balloon during coil placement) provides complete sealing of the inflow zone and durable long-term results.
   - Permanent devices allow reconstruction of the VBJ and preserve anterograde flow to the basilar artery. Treatment of both ruptured and unruptured VBJ aneurysms with stent-assisted coiling and flow-diverter devices (FDD, a flexible stent with low porosity) have been reported.
     - FDDs are mainly used for unruptured aneurysms because the delay necessary to obtain complete occlusion in most cases and the need for antiaggregation therapy and dual antiplatelet therapy makes its use dangerous for ruptured aneurysms.
     - Their use in the setting of an SAH remains the result of a risks/benefits analysis and should be reserved to the otherwise untreatable aneurysms.
     - In large, wide-neck, and fusiform aneurysms, multiple stents and FDDs have been deployed within each other to create a composite endovascular construct and preserve the parent VA.
   - The Pipeline embolization device (Covidien, Mansfield, MA, USA) is currently the only approved FDD in the United States for ICA aneurysms >10 mm. In Europe five types of FDDs have been approved for the treatment of intracranial aneurysms.
   - The reported rates of morbidity (thrombosis, perforators occlusion, intra- and postoperative bleeding), mortality, and recanalization varies considerably between single-center reports; several systematic reviews have been published and meta-analyses are limited by the heterogeneity of data.
   - Complete sealing of the inflow zone is influenced by the size of the device, the number of devices, and the method of deployment.
   - Optimal positioning takes into consideration the location of perforators and branch vessels and the relation between the aneurysm and the loops of the basilar fenestration, if present.
   - In the management of large lesions that do not have a neck, deconstructive techniques can be used (Hunterian ligation aka proximal parent artery occlusion; endovascular trapping preceded with a bypass if needed).
9. *Describe the role of anastomotic revascularization procedures in the management of VBJ aneurysms.*
   - Unclippable and uncoilable VBJ aneurysms may require unilateral VA occlusion.
   - In most patients, unilateral Hunterian VA ligation is well tolerated.
   - When the contralateral VA provides inadequate blood flow, open surgical bypass benefits patients with both ruptured and unruptured aneurysms.
     - This technique (whether intracranial to intracranial bypass or extracranial to intracranial bypass) may lead to cure by trapping complex aneurysms.
     - Anastomotic revascularization procedures require considerable skill and patience.
     - In high-volume cerebrovascular centers, further developments in intraoperative flow assessment, nonocclusive anastomotic technique, graft material, and periprocedural care may improve the efficacy of this high-risk procedure.
10. *You elect a surgical repair via clipping. The procedure is uneventful; however, the patient wakes up with hoarseness and difficulty in swallowing. How do you explain and proceed with this occurrence?*
    - Despite careful handling of the cranial nerves, the patient will often have cranial nerve difficulties postoperatively. These are typically temporary provided that the nerves were preserved and unharmed.
    - Early and aggressive management should be initiated in order to anticipate the risk of aspiration

## Answers (*continued*)

and maximize the likelihood of a favorable recovery. These should include the following:

- Close neuro ICU monitoring:
  - Airway and ventilator support, if necessary
  - Frequent neuro checks
  - Cardiovascular monitoring
  - Basic laboratory evaluation (complete blood count [CBC] to rule out anemia, electrolytes, and coagulation profile)
- Obtain immediate postoperative imaging including CT head and cerebral angiography in order to rule out any potential posterior fossa hematoma, inadvertent injury to perforators, or malposition of the clip. (MRI may be necessary in select cases—clip compatibility should be verified should it be an issue).
- Speech and swallowing evaluation are obtained when the patient is conscious enough including a dynamic barium swallowing study. These may need to be repeated as necessary within a few days to assess patient's progress.
- Initiate IV steroid treatment if cranial nerve manipulation injury is suspected (with gastrointestinal prophylaxis).

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# Case 43 Concomitant Arteriovenous Malformation and Aneurysm

*Anthony M. T. Chau, Cristian Gragnaniello, Remi Nader, Anthony J. Caputy, Dimitri Sigounas, and Marguerite Harding*

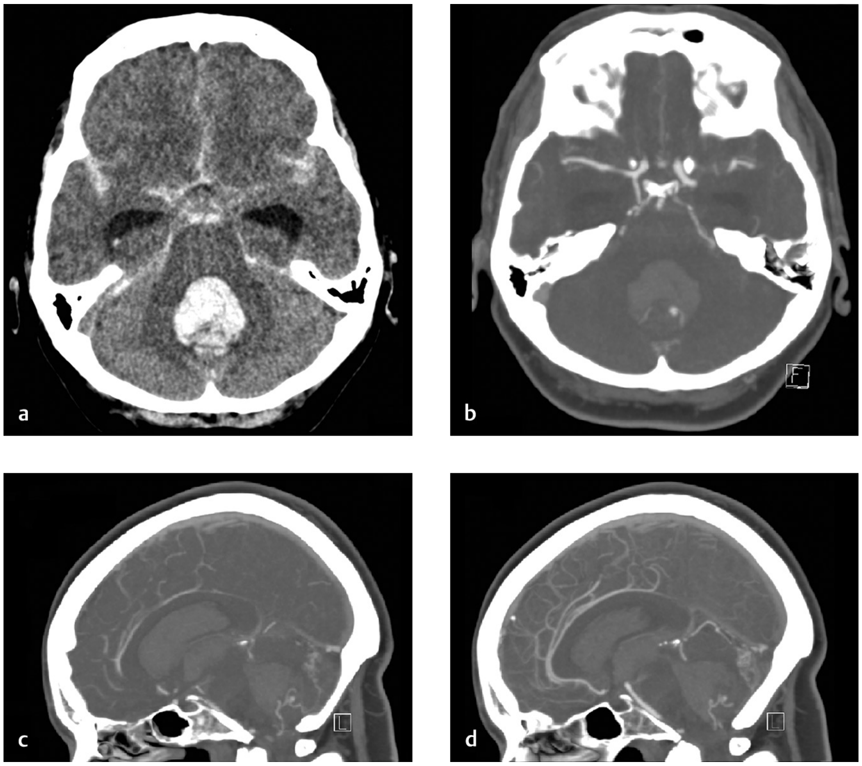

**Fig. 43.1** (**a**) Noncontrast axial CT, (**b**) axial CT angiogram, (**c, d**) sagittal CT angiogram.

## Clinical Presentation

A 35-year-old man with no significant medical history presents with sudden-onset headache and collapse. His pupils are equal and reactive to light. He is intubated and brought to the emergency room. A CT scan and CT angiogram are obtained.

## Questions

1. Describe the CT findings.
2. Describe your initial management.
3. Describe the angiogram findings.
4. What is the correlation between aneurysms and arteriovenous malformations (AVMs)?
5. What is the likely source of the hemorrhage?
6. What are the treatment options?
7. Should the AVM be treated and if so, when and how?
8. How would you manage this patient if he had presented incidentally?

## Answers

1. *Describe the CT findings.*
   - There is diffuse intraventricular and subarachnoid hemorrhage causing obstructive hydrocephalus at the level of the fourth ventricle (▶**Fig. 43.1a**). The pattern of hemorrhage is suggestive of a focus in the posterior fossa. There is no intraparenchymal hemorrhage. There is mild effacement of the posterior fossa cisterns without cerebellar herniation.
   - The CT angiogram demonstrates a cerebellar vermian vascular malformation (▶**Fig. 43.1b–d**). There appears to be a distal left posterior inferior cerebellar artery (PICA) aneurysm.
2. *Describe your initial management.*
   The patient needs to be resuscitated in the emergency room with airway management as well as medical management of blood pressure and intracranial pressure. A frontal ventriculostomy is placed emergently. A four-vessel cerebral angiogram is obtained (▶**Fig. 43.2a**). Further details of management are as per Chapter 38.
3. *Describe the angiogram findings.*
   - There is a grade II Spetzler-Martin (SM) AVM in the cerebellar vermis with a compact nidus less than 3 cm (1 point) in diameter. Its feeders are the bilateral superior cerebellar and PICA arteries. There is deep venous drainage superiorly to the Galenic system (not shown) (1 point). As the cerebellar nuclei and peduncles are not involved, the AVM is not situated in eloquent territory (0 points).
   - There is a large, distal, saccular aneurysm arising midway from the feeding “arterial pedicle” of the left PICA at the apex of a sharp hemodynamic angle (the cranial loop). It is morphologically dysplastic with a daughter saccular outpouching, a 3:1 aspect and 1:1 dome-to-neck ratio. There is no vasospasm or dissection but the PICA distal to the aneurysm is diminished in caliber.
4. *What is the correlation between aneurysms and arteriovenous malformations (AVMs)?*
   - Aneurysms occur in 2% of the general population, but the prevalence is approximately 10 to 20% in patients with AVMs.[1] Increasing age is associated with increasing prevalence of AVM-related aneurysms.[2] The annual rupture rate of cerebral AVMs is 2 to 4%. One study found the rate of hemorrhage at 5 years to be four-fold higher in patients with both AVMs and aneurysms, compared to those with an AVM alone.[3]
   - A handful of AVM-associated aneurysm classification schemes have been proposed. The system by Redekop (1998) classifies aneurysms as intranidal, flow-related, or unrelated to the AVM.[4]
     - Intranidal aneurysms fill early and are located within the AVM nidus.
     - Flow-related aneurysms arise along the course of arteries that go on to supply the AVM. They may be proximal if they are related to the circle of Willis, M1 portion of the MCA or vertebrobasilar trunk, or they may be distal.
     - Unrelated aneurysms occur on arteries that do not supply the AVM.
   - The pathophysiology of flow-related aneurysms (also referred to as feeding artery pedicle aneurysms) is still debated. Coincidental and congenital theories exist; however, the hemodynamic theory has gained most traction in recent times.[1] This theory presupposes that the presence of the AVM produces abnormal stresses on its feeding vessels, predisposing to the formation of aneurysms.
5. *What is the likely source of the hemorrhage?*
   - It can often be difficult to determine the source of bleeding. An institutional series of hemorrhagic presentations reported the source to be the aneurysm in 46% of cases, the AVM in 46%, and uncertain in 8%.[1]
   - The pattern of intraventricular hemorrhage, lack of an intraparenchymal component, combined with the angiographic findings in this case, however, is suggestive of aneurysmal hemorrhage. The aneurysm of the distal left PICA is situated in its telovelotonsillar segment where the bifurcation of the PICA is often located.[5] Rupture of the aneurysm has resulted in blood directed into the fourth ventricle.
6. *What are the treatment options?*
   - Once hydrocephalus and mass effect addressed, management should then be directed at the aneurysm as this is the symptomatic lesion. Definitive treatment of this PICA aneurysm is indicated on an urgent basis. The ideal treatment would involve exclusion of the aneurysm from the circulation while preserving the lumen of the PICA. If this is not possible, as may be the case in dissecting fusiform aneurysms, parent artery occlusion may be required, and the location of the aneurysm relative to proximal brainstem perforators must be carefully considered.
   - The proximal anterior and lateral medullary segments of the PICA invariably give off brainstem perforators, the tonsillomedullary segment may do so occasionally, while it is not the case with the distal telovelotonsillar and cortical segments and their vascular territories may be compensated for by leptomeningeal or anastomotic communications with the superior cerebellar artery (SCA) and anterior inferior cerebellar artery (AICA).[6]
   - Endovascular options for proximal PICA aneurysms include endovascular coiling, at times with the additional need for stent or balloon assistance (▶**Fig. 43.2b**), although this may be associated with higher rates (23%) of aneurysmal recurrence and/or complications.[7] Adequate collateral supply from surrounding cerebellar arteries may allow for safe proximal PICA sacrifice in appropriately selected

## Answers (*continued*)

cases. Distal PICA aneurysms, such as in this case, often present technical difficulties in microcatheter cannulation due to the tortuosity and caliber of the vessel. Vessel sacrifice may be the only endovascular option in some scenarios.

- Surgical strategies include aneurysm clipping, aneurysm trapping with distal bypass/anastomosis (occipital artery–PICA, PICA–PICA), and aneurysmal resection with direct reanastomosis or interposed graft. Lower cranial neuropathy is the main operative concern. The morphology and distal location of the aneurysm make it amenable to direct clip reconstruction via a midline suboccipital craniectomy with the patient in a prone Concorde position, or a left-sided, far lateral craniotomy approach with the patient in the right lateral decubitus position.

7. *Should the AVM be treated and if so, when and how?*
   - A separate decision to treat the AVM needs to be considered. As this AVM has not bled, preference is usually made to address it on an elective basis once the patient has been given an opportunity to recover from the aneurysmal hemorrhage. Even in the scenario of AVM hemorrhage, delaying surgery for 4 to 6 weeks is preferred unless evacuation of a life-threatening hematoma is required.[8] The other major consideration is the patient's age. A crude estimate of percentage lifetime risk of AVM rupture may be given with the following formula: 105 − patient age in years[9]; the formula justifies a more aggressive approach in this 35-year-old, otherwise well patient. For a more detailed estimate, please refer to Case 32: Cerebral Arteriovenous Malformation.
   - An appreciation of the disease's natural history is paramount. Infratentorial AVMs are almost twice more likely than supratentorial AVMs to present with hemorrhage, with concomitantly higher rates of morbidity and mortality.[10] In fact, infratentorial location has been implicated as an independent risk factor for rupture. This may be attributable in part to the poorer tolerance of the posterior fossa feeding arteries to high flow, and the presence of deep venous drainage with complex, hemodynamically unfavorable drainage patterns.
   - This SM grade II (Spetzler-Ponce Class A) AVM would be amenable for surgical resection. This is confirmed with the application of the Lawton–Young supplementary grading scale (2 points for age 20–40 years, 0 points for unruptured, 0 points for compact nidus), which yields a total score of 4 out of a possible 10; 90.9% of such patients can expect a good neurologic outcome following surgical resection.[11] A prospective cohort study demonstrated Spetzler-Ponce Class A unruptured AVMs fared better with surgical resection compared with conservative therapy, even within a short period of follow-up (mean less than 3 years).[12] Preoperative embolization of feeding vessels particularly the SCA in this vermian AVM may be helpful, as the superiorly directed deep venous drainage may obscure intraoperative arterial visualization.[8] Complete resection can be achieved in these AVMs with low morbidity.[8,10]
   - In a mixed population of hemorrhagic and nonhemorrhagic SM I and II AVMs, stereotactic radiosurgery (SRS) used as a primary therapy resulted in a 5-year obliteration rate of 90%, with a median time to obliteration of 30 months, and an annual bleed rate of 2.3%.[13] However, the incidence of post-SRS hemorrhage at the 5-year latency period was some ten-fold higher in patients harboring a coexisting proximal aneurysm (28 vs. 2.6%), resulting in an increased morbidity of 19 versus 1%.[13] This has led some institutions to perform embolization prior to SRS in cases of AVM-associated aneurysms,[14] although this probably results in lower obliteration rates.[15] The role of post-SRS embolization is an avenue of current investigation.
8. *How would you manage this patient if he had presented incidentally?*
   - Patient factors, clinical presentation, and characteristics of the aneurysm and AVM are considered for management of the patient who has presented without hemorrhage. The relationship between flow-related aneurysms and AVMs is imperfectly understood with a paucity of high-quality evidence to guide decision-making. As such, treatment paradigms will vary.
   - In the setting of unruptured pedicle artery aneurysms, some authors have advocated for treatment decisions to be based primarily on the characteristics of the AVM itself.[1] If a decision to address the AVM is made, complete resection is mandated. If both AVM and aneurysm are accessible surgically, then treatment of both at the one sitting may be appropriate. Because primary endovascular or surgical obliteration of the AVM has been observed to result in spontaneous regression of flow-related aneurysms, sole therapy to address the aneurysm has not been recommended by some.[4,16] Others however have advocated aggressive management of flow-related aneurysms before addressing the AVM, citing hemodynamic concerns with abrupt obliteration of the AVM.[17] If SRS is opted for, preradiosurgical endovascular or microsurgical exclusion of the aneurysm should be considered.
   - Overall, an individualized approach is required each time. Acknowledgment of flow-related aneurysms as a significant independent risk factor for future hemorrhage suggests an aggressive approach in the presence of any high-risk features.[18] This would entail an assessment of aneurysmal size, morphology, and growth. In the setting of high-grade AVMs, where conservative treatment may be most appropriate, direct treatment of the aneurysm could be warranted.

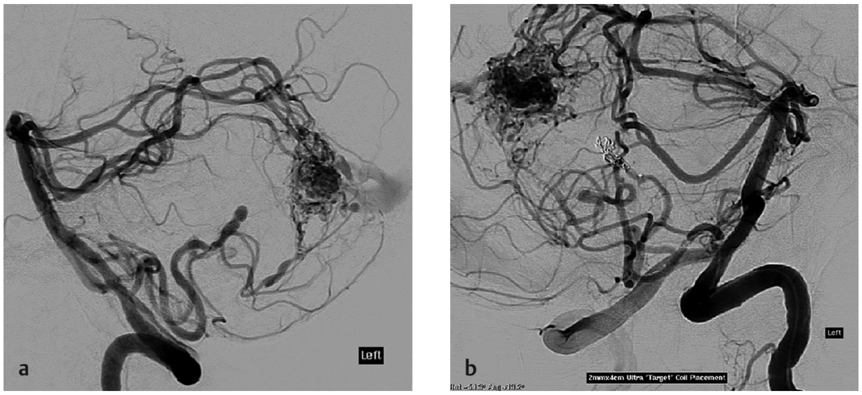

**Fig. 43.2** Digital subtraction angiography, left vertebral artery injection (**a**), lateral view precoiling of left lateral injection of vertebral artery. (**b**) Left oblique view postcoiling.

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# Case 44 Stent- and Balloon-Assisted Coiling

*Layla Batarfi, Mohammed Almekhlafi, and Hosam Al-Jehani*

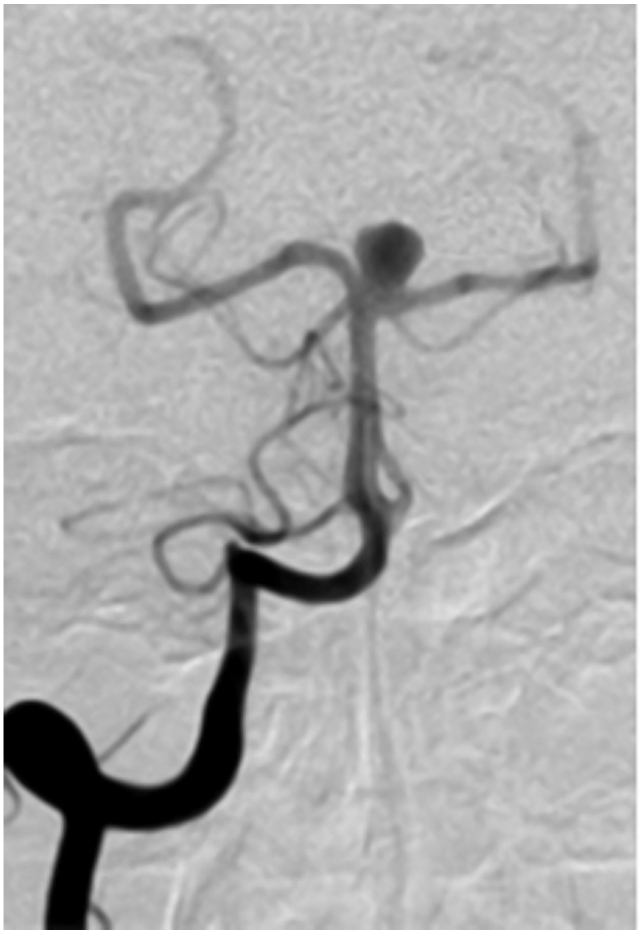

**Fig. 44.1** CT angiogram.

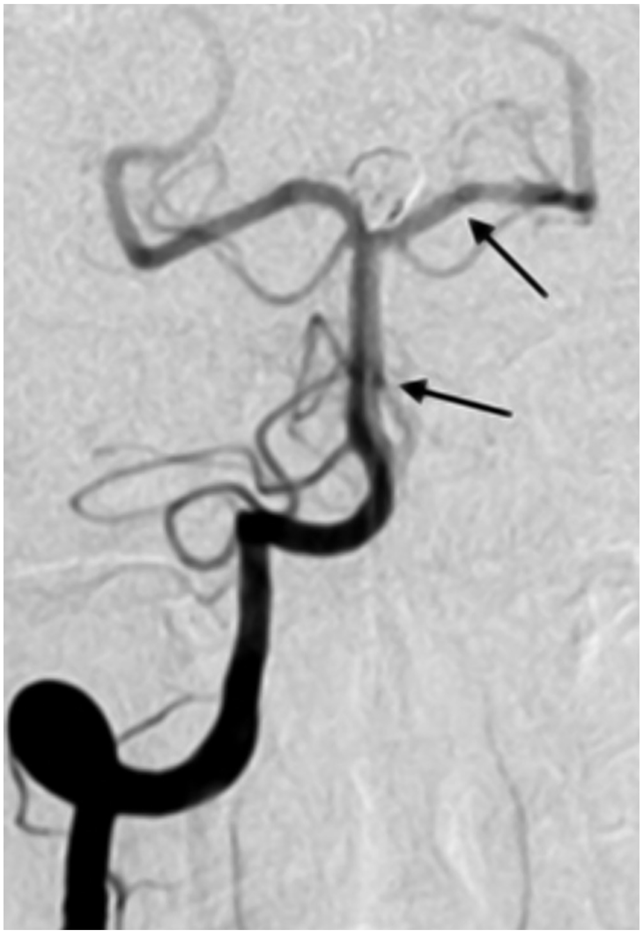

**Fig. 44.2** Postprocedural angiogram.

## Clinical Presentation

- A 43-year-old male previously healthy suddenly developed severe unrelenting headache while working out.
- He was found to be neurologically intact with a CT scan showing subarachnoid hemorrhage (SAH) mostly in the interpeduncular cistern and extending to the prepontine cistern and both Sylvan fissures with no intraventricular extension and no hydrocephalus.
- Angiogram was done, which revealed a single pertinent finding (see ▶**Fig. 44.1**).

## Questions

1. Describe the angiogram.
2. What is the diagnosis?
3. What would be considered as a “wide-neck” intracranial aneurysm?
4. Outline a general management for patients with giant aneurysm with SAH.
5. If clipping is not available or not possible, describe the endovascular techniques available to treat this lesion.
6. Describe the postprocedural angiogram (▶**Fig. 44.2**).

## Questions (*continued*)

7. If the aneurysm is too complex for standard assisted coiling what are your options?
8. What are the common risks associated with the endovascular management of aneurysms?

## Answers

1. *Describe the angiogram.*
   - This is an anteroposterior view of a right vertebral artery injection showing a wide-neck aneurysm the origin of the left posterior cerebral artery (PCA).
   - The aneurysm is pointing superiorly and has a slightly irregular dome appearance.
2. *What is the diagnosis?*
   - Aneurysmal SAH, World Federation of Neurological Surgeons (WFNS) grade 1, secondary to the rupture of a wide-neck left PCA aneurysm.
3. *What would be considered as a "wide-neck" intracranial aneurysm?*
   - Traditionally, any aneurysm with a neck larger than 4 mm was considered wide necked. Then several geometric determinants were studied and the aspect ratio or a dome-to-neck ratio was found to be of significant and a cutoff of less than a factor of 2 was considered as the limit for wide-neck aneurysms.
   - These measures were devised when the technology of available interventional material was at its beginnings.
   - More recently, it can be derived from the literature that aneurysms with aspect and dome-to-neck ratios > 1.6 usually did not require adjunctive techniques, while aneurysms with aspect and dome-to-neck ratios < 1.2 almost always required adjunctive techniques.[1,2]
4. *Outline a general management for patients with giant aneurysm with SAH.*
   - Admit to intensive care unit (ICU) for close monitoring of vital signs and close hourly follow-up via neurological exam.[3,4]
   - Activity level limited to bed rest with head of bed elevated at 30 degrees.
   - SAH precautions (i.e., low level of external stimulation, restricted visitation, no loud noises)
   - Recommended diet: NPO (in preparation for surgery or endovascular intervention)—intravenous (IV) fluids: early aggressive fluid maintenance therapy
   - Medications
     - Prophylactic anticonvulsants
       - Controversial but typically given for at least for 1 week postoperatively
       - Options include Keppra (levetiracetam, start with 500 mg PO or IV q 12 hours) and phenytoin may be used (load with 17 mg/kg, maintenance of 300 mg daily)
     - Sedation for agitation: for example, with propofol or dexmedetomidine
     - Analgesics: fentanyl. It has the advantage of lowering intracranial pressure (ICP; 25–100 mg q 1–2 hours PRN)
     - Dexamethasone (Decadron): controversial; not as an acute measure, may help with headache and neck pain related to chemical meningitis from the SAH
     - Stool softener in awake patients
     - Antiemetics: ondansetron 4 mg IV q4 hours PRN
     - Nimodipine (Nimotop) 60 mg PO/NG q 4 hours initiated within 96 hours of SAH, IV administration is equally effective. Mainly as a neural protection at the time of onset of vasospasm
     - $H_2$ blockers or proton pump inhibitors to reduce risk of stress gastric ulceration
     - Tight blood pressure control: systolic blood pressure (SBP) 120 to 150 mm Hg by cuff is a guideline with unsecured aneurysms
   - Laboratory studies:
     - Complete blood count (CBC), electrolytes, prothrombin Time/partial thromboplastin time (PT/PTT), arterial blood gas (ABG) and electrocardiogram (ECG) on admission
     - ABG, electrolytes, CBC q day or as necessary depending on patient status
     - Osmolality: serum and urine, if urine output high or low to distinguish cerebral salt wasting from syndrome of inappropriate antidiuretic hormone (ADH) in the context of hyponatremia
     - Chest X-ray (CXR) as needed, if risk of pulmonary edema is present
5. *If clipping is not available or not possible, describe the endovascular techniques available to treat this lesion.*
   - Balloon-assisted coiling (BAC):
     - This involves placement of a removable balloon adjacent to the aneurysm. This prevents coil loops from herniating into the parent vessel and helps the coil to assume the shape of the aneurysm.[5]
     - With the inflation of a balloon, systemic heparinization has to be initiated. The inflations should be limited to 3 or 5 minutes during which no more than one coil can be deployed.
     - If upon deflation, the coil mass is integrated well within the aneurysm the procedure can be concluded.
     - However, in case loops of coil prolapse or herniate into the parent vessel, then there are two options:
       - Deploy more coils for better packing of the aneurysm leading to the formation of a tighter mesh of coils.

## Answers (*continued*)

    - ○ Deploy a stent across the neck to prevent the herniation of the coil loops (see below).
  - Stent-assisted coiling (SAC)
    - This involves permanent placement of a stent in the vessel adjacent to the aneurysm to provide a scaffold that improves aneurysm neck coverage, thus allowing optimal aneurysm coiling.
    - This could be done by either the "jailing" technique in which the coiling microcatheter is passed into the aneurysm first then the stenting microcatheter is passed and the stent is deployed followed by coiling then withdrawal of the "jailed" microcatheter upon achieving good coil packing.
    - Alternatively, this can be done by deploying the stent first, followed by passing the microcatheter through the pores of the stent and completing the coiling through the stent.[6]
    - It has to be emphasized that with any stenting procedure the patient has to be properly prepared with dual antiplatelets with verification of response before stent deployment.
    - If the patient is a hypo responder or the need for the stent becomes realized only during the procedure with no neurosurgical expertise available, then the patient can be loaded with dual antiplatelets on the angiographic table and the stenting can then be undertaken. The patient should continue dual antiplatelets for 3 months followed by a single agent for life, to prevent in-stent thrombosis.

6. *Describe the postprocedural angiogram (▶* **Fig. 44.2***)*
   - This is the same anteroposterior projection of the right vertebral artery. It shows the complete packing of the aneurysm via coiling.
   - The arrows point to the proximal and distal limits of the stent deployed across the neck of the aneurysm to protect the P1 segment form coil herniation and occlusion.
7. *If the aneurysm is too complex for standard assisted coiling what are your options?*
   - Surgical clipping is always a viable option either alone or with wrapping and bypass.
   - Partial clipping can be considered in order to modify the aneurysm enough to render it amenable to coiling or assisted coiling. Same can be said about partial coiling in the acute setting of the SAH then when the patient is stabilized, the decision is made as to the final obliteration option.
   - Parent vessel occlusion in the presence of good cross-flow verified by test balloon occlusion. This can be done either surgically or via endovascular approaches.
   - The use of telescoping stents, overlapping at the area of the neck can be considered in order to increase coverage across a very wide-neck difficult aneurysm.
   - Flow-diverting stents can be used in the treatment of unruptured or ruptured large wide-neck aneurysms or saccular ones occurring along a side branch, when other nonsurgical treatment options cannot be performed.[7]
8. *What are the common risks associated with the endovascular management of aneurysms?*
   - Thromboembolic events
     - This is a major concern. Reports vary widely, with estimated rates ranging from 2.5 to 28%
     - This notoriously higher risk is thought to be associated more with undergoing balloon-assisted procedures
   - Hemorrhagic complications
     - This is related to the early and prolonged use of dual antiplatelets. New stents are being developed to overcome this need, but they are not in wide use yet.
     - The use of dual antiplatelets is associated with a marginal increase in the incidence of hemorrhagic events, but it unfortunately leads to a significant increase in severity of the hemorrhage, should it occur, with a much worse morbidity and mortality profile.
   - Intraprocedural rupture
     - Cited in most studies to be between 2 and 8% causing morbidity or mortality in up to 50% of patients
     - Risk factors for intraprocedural rupture include small aneurysm size, recent rupture, and presence of a daughter sac
   - Recanalization
     - SAC showed significantly better results than the standalone coiling as major recurrences were observed in 9.7% of patients treated with standalone coiling compared with 3.9% in the stent group.

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# Case 45 Balloon Test Occlusion and Giant Aneurysm

*Mohammad A. Aziz-Sultan*

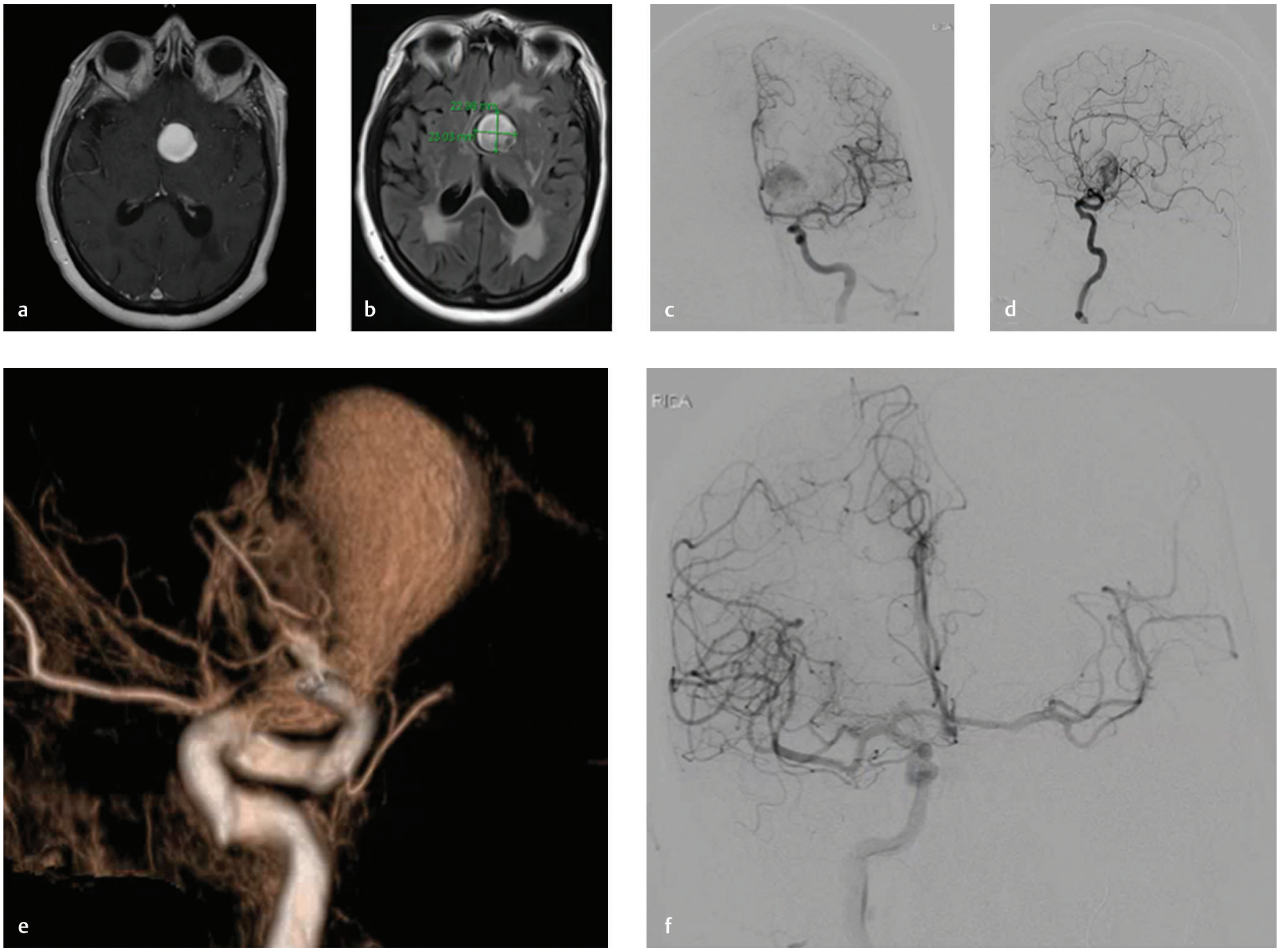

**Fig. 45.1** Relevant MRI axial views T1-weighted sequence postcontrast (**a**), T2-weighted sequence (**b**). Cerebral angiogram left internal carotid artery (ICA) injection; anteroposterior view (**c**), lateral view (**d**), and 3-D reconstruction (**e**). Balloon test occlusion angiogram right ICA injection; anteroposterior view (**f**).

## ■ Clinical Presentation

- A 72-year-old woman with a history of hypertension, hypothyroidism, thrombocytopenia, anxiety, and intracranial aneurysm presents to the emergency department after developing word finding difficulty and short-term memory loss.
- She denied any acute-onset headaches, nausea, emesis, dizziness, falls, focal motor or sensory deficits, and incontinence.
- The patient was being followed up for partially thrombosed calcified aneurysm and when compared with previous imaging, the aneurysm has enlarged.
- New MRI and angiogram obtained, pertinent images presented in ▶**Fig. 45.1**.

## Questions

1. Describe the findings on the images (▶**Fig. 45.1**).
2. What type of aneurysm does this patient have? What is the typical presentation for these aneurysms?
3. What are some treatment options for this patient?
4. What is a balloon test occlusion (BTO) procedure? What information does this procedure provide in this case?
5. What are some indications for BTO? What are some special considerations when performing BTO?
6. What are the major components of a BTO exam?
7. What is the predicative value of BTO?

## Answers

1. *Describe the findings on the images* (▶**Fig. 45.1**).
   - MRI of the brain (axial T1-weighted with contrast and axial T2-weighted images) shows a partially thrombosed calcified aneurysm measuring approximately 2.8 × 2.9 cm.
   - There appears to be significant mass effect on the third ventricle with subsequent enlargement of both lateral ventricles, left greater than right.
   - There is evidence of transependymal edema indicative of obstructive hydrocephalus.
   - Digital subtraction angiography (DSA) shows a giant left ophthalmic segment internal carotid artery (ICA) aneurysm with a superiorly and medially projecting dome measuring 28 mm in greatest height seen to fill in a delayed fashion and exhibiting contrast stagnation.
   - The anterior cerebral arteries are shifted from left to right secondary to mass effect.
   - Additional temporal information: an angiographic venous delay of 6 seconds was noted between the two hemispheres during right internal carotid angiography with left internal carotid balloon occlusion. No significant retrograde flow across the left posterior communicating artery was seen upon left vertebral angiography during left internal carotid balloon occlusion. Taken together, these findings were indicative of poor collateral flow to the left hemisphere in the face of left carotid occlusion.
2. *What type of aneurysm does this patient have? What is the typical presentation for these aneurysms?*
   - This patient has a left partially thrombosed giant ophthalmic segment ICA aneurysm.
   - Unruptured aneurysms are typically assessed based on size and location in terms of risk for subarachnoid hemorrhage (SAH).
   - Based on the International Study of Unruptured Intracranial Aneurysms Investigators (ISUIA) findings, this patient has a giant aneurysm defined as > 24 mm in the anterior circulation with an annual risk of rupture of 8% (▶**Table 45.1**).
   - Giant aneurysms account for ~5% of all intracranial aneurysms, slight female predominance, and present in the fifth to seventh decades of life.[1]
   - Approximately 30 to 50% patients present with SAH and 50 to 70% have luminal thrombus with surrounding edema.[1–3]
   - Due to this thrombotic phenomenon, ~8% will sustain an embolic event.[1]
3. *What are some treatment options for this patient?*
   Giant aneurysms may be treated in various ways, through multiple modalities. Some treatment options considered for this patient include the following:
   - Open surgical:
     - Clipping
     - Trapping +/– bypass
     - Wrapping
   - Endovascular:
     - Coiling
     - Stent-assisted coiling
     - Flow diversion
     - Parent artery ligation/occlusion
       For this patient, the parent artery occlusion/ligation and bypass was the treatment selected by the authors.
4. *What is a balloon test occlusion (BTO) procedure? What information does this procedure provide in this case?*
   - When considering parent artery ligation, therapeutic occlusion, in treatment of giant aneurysms BTO test must be performed to asses for potential strokes or deleterious effects of vessel sacrifice.
   - Blind carotid artery sacrifice without any temporary test occlusion may result in stroke in approximately 17 to 30% of individuals.[5–9]
   - First described by Serbinko in the 1970s, BTO has remained relatively unchanged.[10] This involves introduction of a luminal balloon catheter that is inflated in the desired vessel to the point of flow arrest in a conscious patient.
   - There have been multiple adjunctive tests since its first inception to improve BTO prognostic value.
   - In principal, BTO allows one to assess intracranial collateral circulation during temporary occlusion of a diseased vessel (▶**Table 45.2**).
5. *What are some indications for BTO? What are some special considerations when performing BTO?*
   - Indications for BTO include:
     - Giant intracranial aneurysms

## Answers (*continued*)

  - Pseudoaneurysms of the ICA or vertebral artery
  - High-risk procedures involving intracranial vessels or their parent arteries
  - Neoplasms involving carotid arteries
  - Carotid-cavernous fistula or arterial dissections when anticoagulation is contraindicated, and acute/active extravasation
- Special consideration must be made during BTO to perform temporary occlusion as close as possible to the intended point of ligation, in particular, when considering ligation of ICA for aneurysms.
- Patients with retrograde filling through an ophthalmic artery may show a false negative test when performed proximal to the origin of the ophthalmic artery; this can be seen in up to 14% of patients.[11]
- In the paraophthalmic ICA, distal to the ophthalmic artery, BTO is recommended with visual testing.[12]
- Conversely, in highly vascular neoplasms care must be taken to perform BTO distal to the tumor in order to prevent steal phenomena from altering BTO results.[13]
- BTO and point of temporary occlusion should be catered to the specific patient needs and site of intended permanent occlusion.

6. *What are the major components of a BTO exam?*
- BTO is usually performed through femoral access with a 6-French sheath.
- The desired vessel is then accessed through a 5-French guide catheter and positioned proximal to the area of interest.
- Over roadmaps and fluoroscopy, an appropriately sized balloon catheter (▶**Table 45.3**) is placed at the point of potential occlusion, avoiding tortuous or atherosclerotic segments.
- Once in position, a balloon is inflated slowly to the point of flow arrest.
- Once this is achieved, timing begins and the exam is carried out for a minimum of 15 minutes, but not more than 30 minutes. Clinical and adjunct testing maybe performed at this time.
- It is our practice to perform clinical exams and two adjunctive tests:
  - Clinical exam with hypotensive challenge:
    - Clinical exam is performed at baseline and at 5-minute intervals during the procedure.
    - Careful note is made regarding any alterations in mental status, acute neurological deficits, and the development of nausea, anxiety, or agitation.
    - This can be supplemented with neuropsychological testing to assess higher cortical function. Although this further detects subtle findings of dysfunction, it requires the aid of skilled personnel for administration and assessment.
    - In addition, the mean arterial pressure (MAP) can be lowered ~65% of baseline to further stress the collateral flow and altered hemodynamics during BTO.
    - Clinical examination should be performed for a further 10 to 15 minutes with induced hypotension.
    - If deficit develops or patient becomes intolerant of hypotensive challenge, then the procedure should be aborted and the territory is considered at risk for ischemia.
    - For this reason, short-acting and readily reversible agents, such as nitroprusside and esmolol, should be utilized.
    - Although hypotensive challenge can add to BTO and further delineate patients with at-risk territory, there remains a significant subset of patients that may develop delayed cerebral ischemia with a false negative rate of up to 15%.[14]
  - Angiography:
    - A formal angiogram is preformed prior to parent vessel occlusion.
    - Careful note is made of anastomotic vessels and the circle of Willis, and venous phases.[15]
    - Once the balloon has been inflated and the parent vessel has been occluded, repeat angiography is performed.
    - Again, note is made of cross-filling and collateral opacification of territory at risk.
    - The venous phase is also carefully studied for synchronicity of venous filling.
    - The timing of venous phases within 2 second between the occluded and nonoccluded sides has been correlated with uneventful parent artery ligation.[16]
  - Single-photon emission computed tomography (SPECT):
    - Radioactive tracers can further aid in determining areas at risk for ischemia.[17]
    - 99m-technetium-hexamethylpropyleneamine oxime (HMPAO) is injected 5 minutes after initiating occlusion.
    - The tracer then deposits into brain tissue in proportion to cerebral blood flow.
    - While most imaging studies would necessitate transport during the procedure, SPECT has the added benefit of being performed after completion of BTO.
    - After completion of the BTO, a SPECT scan can be performed 1 to 6 hours later.
    - This study adds highly sensitive qualitative analysis of impaired cerebral blood flow.
    - The success of carotid artery sacrifice in the setting of a negative BTO and adjunct SPECT approaches 100%.[18]

## Answers (*continued*)

- At the end of the test the balloon is deflated, all catheters are removed, and the arteriotomy is closed with Angio-Seal or manual compression with hemostasis.

7. *What is the predicative value of BTO?*
    - Although BTO reduces the risk of postoperative stroke, there still remains chances of 5 to 40% stroke rate (R.10–11).
    - This risk can be significantly reduced with the addition of adjunct testing.
    - Standardized method and angiography with careful attention to venous phase delay can yield a positive predictive value of up to 98%.[19]
    - Cerebral blood flow is preserved in these patients with synchronous venous filling on BTO in the long term.[20]
    - Careful evaluation of clinical examination and angiographic data, including synchronicity of venous phase, with adjunctive testing can significantly increase the positive predictive value of BTO.
    - BTO itself is a relatively safe procedure with a complication rate of about 3.2% and can guide the treatment of cerebrovascular pathology.[21]

**Table 45.1** ISUIA findings of annual rupture rates (%) according to size and location[4]

| Aneurysm size | Anterior circulation (anterior communicating artery/MCA/ICA) | Posterior circulation (posterior communicating, basilar, and vertebral arteries) | Cavernous ICA |
|---|---|---|---|
| < 7 mm + history of SAH | 0 | 0.5 | 0 |
| < 7 mm − history of SAH | 0.3 | 0.68 | 0 |
| 7–12 mm | 0.52 | 2.9 | 0 |
| 13–24 mm | 2.9 | 3.68 | 0.6 |
| > 24 mm | 8.0 | 10 | 1.28 |

*Abbreviations:* ICA, internal carotid artery; ISUIA, International Study on Unruptured Intracranial Aneurysms; MCA, middle cerebral artery; SAH, subarachnoid hemorrhage.

**Table 45.2** Risk of parent artery ligation and results of BTO

| Risk of Parent Artery Ligation | Clinical Exam with Hypotensive Challenge | Angiography with Venous Phase | Intervention |
|---|---|---|---|
| Low risk | Pass | Pass | Parent artery occlusion |
| Moderate risk | Pass | Fail | Consider low-flow bypass or alternative treatment |
| High risk | Fail | Fail | Consider high-flow bypass or alternative treatment |

*Abbreviation:* BTO, balloon test occlusion.

**Table 45.3** BTO and catheter selection

| Vessel to undergo BTO | Balloon catheter (mm) | Average vessel size/ caliber (mm) |
|---|---|---|
| Cervical ICA | 7 × 7 HyperForm | 5 |
| Intracranial ICA | 4 × 7 HyperForm | 3.5 |
| Vertebral artery | 4 × 10 Hyperglide | 4 |
| Basilar artery | 4 × 7 HyperForm | 3.2 |

*Abbreviations:* BTO, balloon test occlusion; ICA, internal carotid artery.

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# Case 46 Ischemic Stroke Initial Management

*Shaymaa Al-Umran, Mohammad Almekhlafi, and Hosam Al-Jehani*

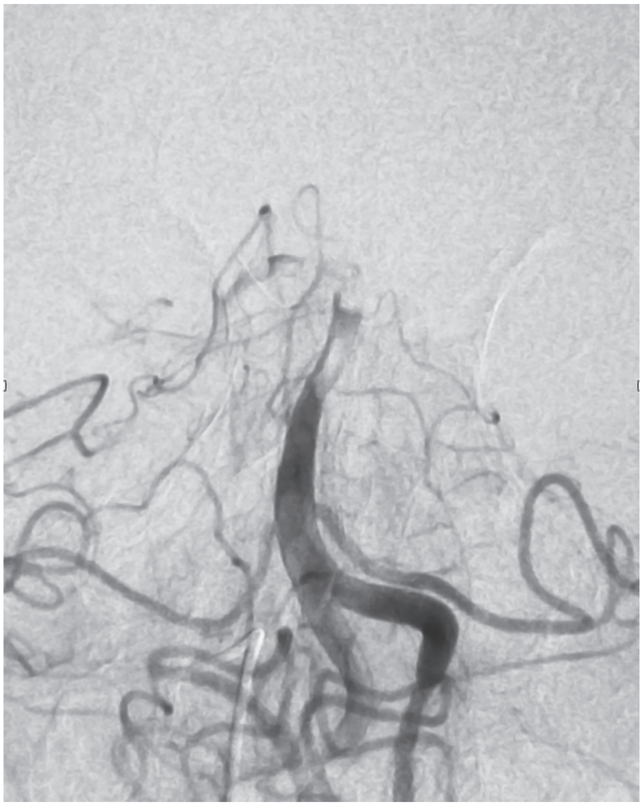

**Fig. 46.1** Cerebral angiogram—anteroposterior projection of the left vertebral artery.

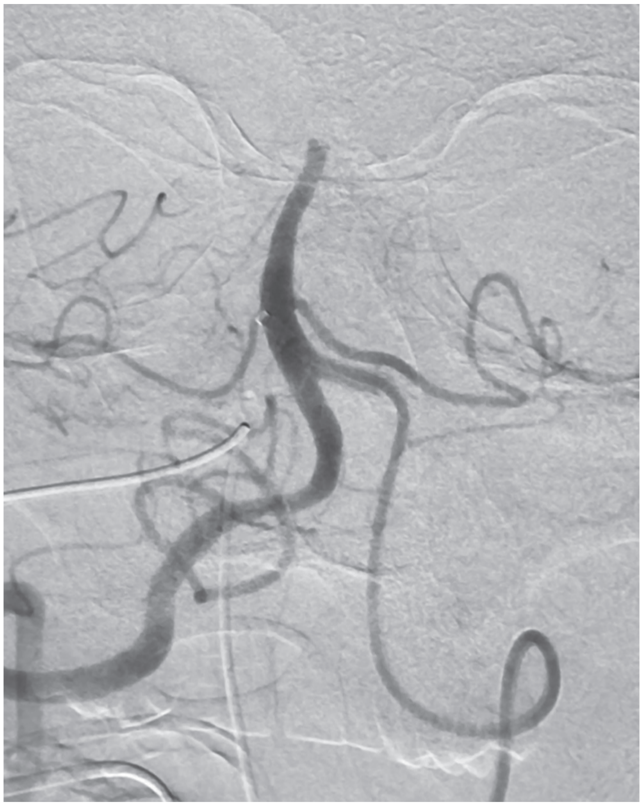

**Fig. 46.2** Cerebral angiogram—anteroposterior projection of the left vertebral artery, post stent retrieval attempt.

## Clinical Presentation

- A 47-year-old hypertensive male presents with sudden onset of dizziness and mild left-sided clumsiness and weakness.
- While being worked up, he suddenly collapsed to a Glasgow Coma Scale (GCS) of 4/15 with motor weakness noted on the left side. An urgent CT scan showed no gross abnormalities.
- The suspicion of a vascular event yielded the acquisition of a CT angiogram/angiogram showing distal basilar occlusion.

## Questions

1. Describe the initial angiogram (▶**Fig. 46.1**).
2. What are the indications for intravenous (IV) tissue plasminogen activator (t-PA)?
3. What are the contraindications to IV t-PA?
4. Are there any other considerations that could alter the timing of the interventions for acute ischemic strokes?
5. What is the success rate of IV t-PA?
6. How would vascular imaging help the decision-making in cases of acute stroke?
7. Discuss the role and timing of endovascular therapy.
8. How do posterior circulation stroke differ from anterior circulation stroke?
9. What is the time line for acute posterior circulation stroke intervention?

   The patient was submitted to stent retrieval attempt, passing an appropriate-sized stent into the clot three times; each attempt lasting 7 minutes. This angiogram was obtained after the third pass (▶**Fig. 46.2**).
10. Describe the postprocedural angiogram (▶**Fig. 46.2**).

## Answers

1. *Describe the initial angiogram.*
   - This is an anteroposterior projection of the left vertebral artery injection showing a distal basilar occlusion with no visualization of either superior cerebellar arteries or the posterior cerebral arteries.
   - A long filling defect is noted in the mid to distal basilar artery before it is complete cut off, suggesting a long intra-arterial thrombus.
2. *What are the indications for intravenous (IV) tissue plasminogen activator (t-PA)?*
   - Indications include:[1]
     – Time since last seen with a normal neurological examination < 4.5 hour
     – Age ≥ 18 years
     – Diagnosis of ischemic stroke causing disabling neurologic deficit
3. *What are the contraindications to IV t-PA?*
   - Clinical:[1]
     – Sustained hypertension above 185/110 mm Hg
     – Symptoms suggestive of subarachnoid hemorrhage
     – Previous history of intracranial hemorrhage (ICH)
     – ST-elevation myocardial infarction within the previous 3 months
     – Major head trauma or stroke within the previous 3 months
     – Major surgery within the previous 14 days
     – Gastrointestinal or urinary tract hemorrhage within the previous 21 days
     – Arterial puncture at a noncompressible site within the previous 7 days
     – Recent lumbar puncture
     – Active bleeding or acute traumatic fracture on examination
     – Seizure at onset with suspected postictal deficits
     – Minor or rapidly improving neurologic deficits
     – Known intracranial aneurysm or arteriovenous malformation (AVM; caution)
   - Radiologic:
     – Head CT showing hemorrhage or multilobar infarction.
   - Laboratory:
     – Oral anticoagulation with international normalized ratio (INR) > 1.7
     – Heparin administered within the previous 48 hours with elevated current activated partial thromboplastin time (aPTT)
     – Platelet count < 100,000 per mm$^3$
     – Blood glucose level greater than 400 mg/dL or less than 50 mg/dL (2.7 mmol/L) at presentation with improving deficits following correction of hypoglycemia
4. *Are there any other considerations that could alter the timing of the interventions for acute ischemic strokes?*
   - Contraindications for treatment between 3 and 4.5 hours according to European Cooperative Acute Stroke Study (ECASS) III criteria:
     – Age > 80 years
     – Very severe deficits at onset (National Institutes of Health Stroke Scale [NIHSS] score > 25)
     – Combination of previous stroke and diabetes mellitus
     – Prior warfarin therapy irrespective of the INR
5. *What is the success rate of IV t-PA?*
   - This highly depends on the vessels occluded and whether vessels proximal or distal to the thrombus are occluded as well.
   - Overall recanalization rate is 8 to 10%.
6. *How would vascular imaging help the decision-making in cases of acute stroke?*
   - Brain imaging is the cornerstone of evaluation.
   - Findings to consider include the size, location, and vascular territory of the infarction, the presence of intracranial bleeding, or the presence of acute complications such as hydrocephalus or a mass.[2]
   - CT:
     – Noncontrast CT scan of the head is sufficient to exclude ICH, and emergency treatment should not be delayed to obtain more advanced imaging modalities such as MRI.
     – Early signs of brain ischemia can often be seen when the CT scan is assessed in detail, these may be as follows:
       ○ Hyperdense middle cerebral artery sign is associated with worse prognosis and higher risk of hemorrhage after fibrinolysis, but IV recombinant (rt)-PA can still be useful for these patients.
       ○ Loss of insular ribbon
       ○ Obscuration of the lenticular nucleus
       ○ Loss of gray-white matter differentiation and focal sulcal effacement
   - CT angiography (CTA):
     – The primary goal of CTA is to identify patients who have a major arterial occlusion that may not respond well to intravenous thrombolysis or may portend risk of a large hemispheric infarction.
   - CT perfusion:
     – Usually obtained with CTA
     – Helps identify tissue that may be ischemic, but not yet infarcted (ischemic penumbra versus a well-established nonsalvageable infarction)
     – Allows the measurement of three parameters: cerebral blood volume (CBV), mean transit time (MTT), and cerebral blood flow (CBF). Mismatch between CBV/CBF and MTT indicates salvageable penumbra.
     – Useful in wake-up strokes where time of onset cannot be determined
   - MRI:
     – MRI can also be useful for the emergency evaluation of patients with suspected stroke.

## Answers (*continued*)

  - Advantages:
    - Detection of acute, small, and posterior fossa infarctions
    - Differentiation of acute from older infarction
    - Excellent spatial resolution for tissue edema
    - Perfusion sequences can assess the time to peak, MTT, CBF, and CBV allowing for detailed assessment of penumbra.
    - For added validity, comparison of the volume of the lesions found between diffusion-weighted imaging (DWI) and perfusion sequences can be used to better delineate the ischemic penumbra.
  - Disadvantages:
    - Time delays
    - Cost
    - Relatively limited availability
    - Contraindications including:
      - Claustrophobia
      - Cardiac pacemakers and ferromagnetic metal implants
      - Nephrogenic systemic fibrosis has been associated with gadolinium used for enhanced MRI studies, and patients' renal function should be screened before the agent is given. This can be mitigated by the use of time-of-flight MR angiography (TOF MRA).
      - Pregnancy is a contraindication for gadolinium injection
- MRA:
  - This can demonstrate severe stenosis or occlusion of the major intracranial or extracranial vasculature.
- Transcranial Doppler (TCD):
  - It is commonly used in acute stroke evaluation to diagnose a major cerebral artery occlusion that may dictate advanced treatment.
  - Examples of such decision include cerebral sickle crises is response to thrombolytic +/– US thrombolysis.

7. *Discuss the role and timing of endovascular therapy.*
   - Current guidelines indicate that intra-arterial thrombolysis is an option for the treatment of patients within 6 hours of a major stroke, secondary to an occlusion of the middle cerebral artery, who cannot be treated with IV t-PA.
   - This time window can be expanded to 8 to 24 hours by use of stent retrieval device, as long as the imaging is revealing a salvageable penumbra.[3–5]
   - Recent studies favor the rapid institution of endovascular intervention in acute ischemic stroke with large proximal vessel occlusion to promote rapid recanalization and boost good collateral circulations.[3–5]
8. *How do posterior circulation stroke differ from anterior circulation stroke?*
   - Posterior circulation stroke differs in from anterior circulation stroke in several aspects:
     - The evolution of clinical symptoms is often gradual, making precise assessment of the onset of symptoms and the time window for treatment difficult.
     - Atherothrombosis (unstable plaque with thrombus) is more common than embolic events in posterior circulation strokes.
     - The risk of reocclusion after recanalization is high.
     - The natural history shows a poor outcome with a high mortality rate of 70 to 80% unless recanalization is achieved.
     - Most patients who present with complete occlusion of bilateral vertebral arteries or the basilar artery have acute, severe, and often life-threatening strokes at the time of occlusion. However, some patients survive with chronic bilateral vertebral artery occlusion because they develop collaterals through posterior communicating arteries that are present but inadequate or through other unusual collateral pathways such as the anterior spinal artery.
9. *What is the time line for acute posterior circulation stroke intervention?*
   - Timeline for IV t-PA is not different and limited by 4.5 hours from onset of deficits.
   - Depending on preservation of brainstem function, the window for stent retrieval (+/–thrombolysis) can be extended up to 36 to 48 hours depending on the center of practice.
   - It is highly advisable to obtain an MRI to assess the viability of the brainstem before committing to such aggressive measure.[6]

   The patient was submitted to stent retrieval attempt, passing an appropriate-sized stent into the clot three times; each attempt lasting 7 minutes. This angiogram was obtained after the third pass (▶ **Fig. 46.2**).
10. *Describe the postprocedural angiogram.*
    - This is the same projection of the left vertebral artery injection. It shows the persistence of the distal basilar occlusion, with disappearance of the previously seen filling defect in the mid basilar artery.

    At this point, a dedicated suction (intermediate) catheter was introduced at the base of the visualized clot (under road-map guidance) and the clot was successfully aspirated.[7] The postaspiration angiogram shows complete filling of the superior cerebellar arteries and both posterior cerebral arteries (▶ **Fig. 46.3**). The patient was extubated the next morning with a GCS of 15 and stable mild left hemiparesis.

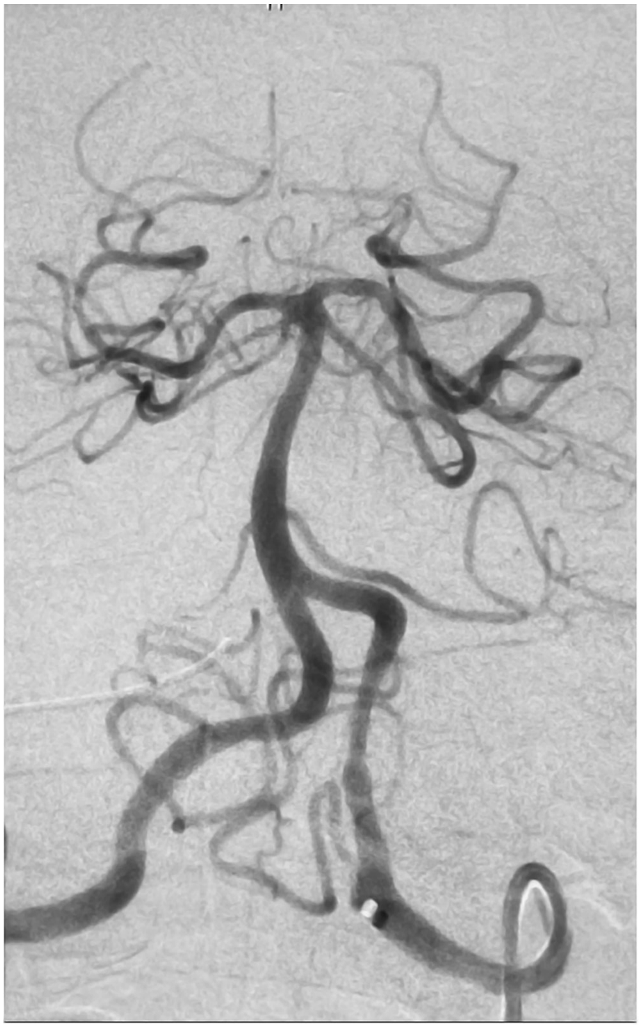

**Fig. 46.3** Cerebral angiogram—anteroposterior projection of the left vertebral artery, post suction catheter.

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# Case 47 Decompressive Craniectomy for Ischemic Stroke

*Justin Reagan, Alan Siu, Cristian Gragnaniello, Dimitri Sigounas, Anthony J. Caputy, and Zachary N. Litvack*

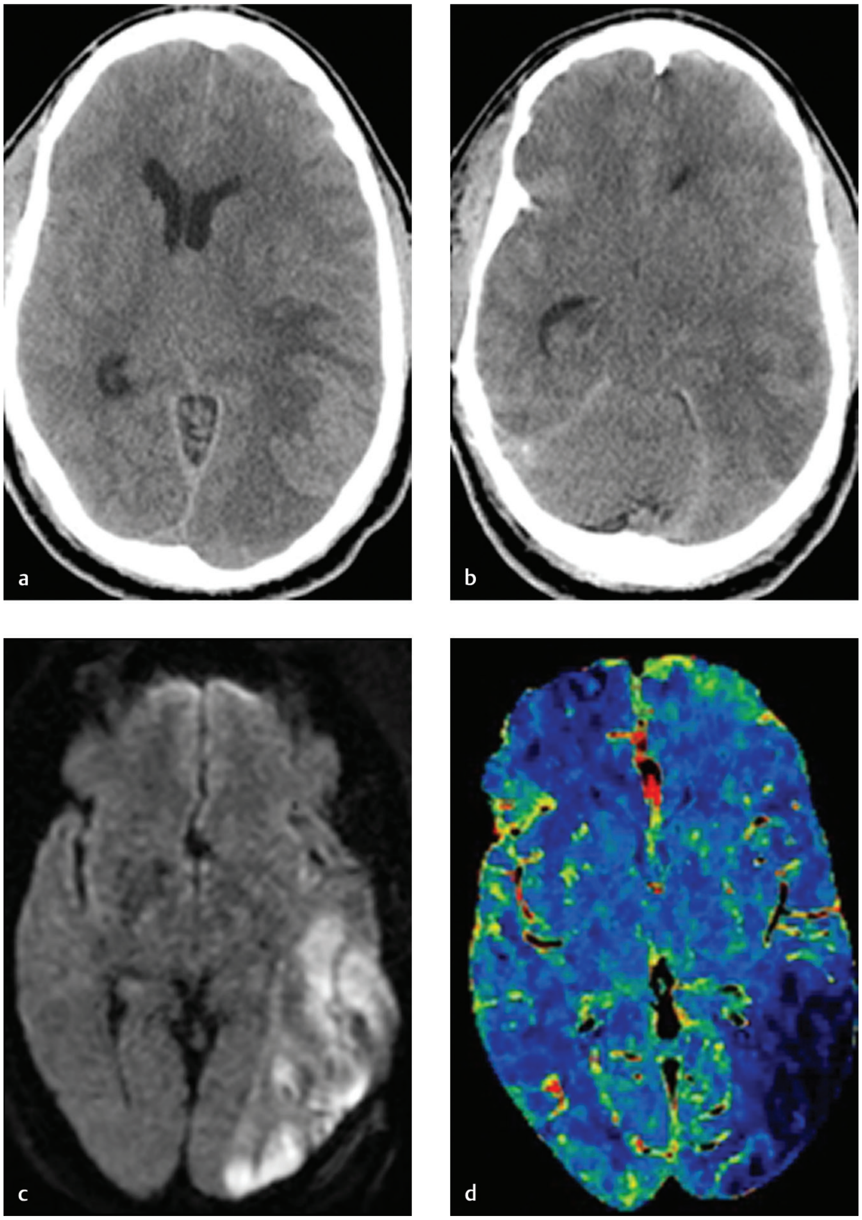

**Fig. 47.1** CT scan without contrast at the level of the ventricles (**a**) and floor of the middle fossa (**b**) demonstrating effacement of the basal cisterns, midline shift, and left occipitotemporal malignant cerebral edema. MRI of cerebral blood flow (**c**) and diffusion-weighted imaging (**d**) revealed a left occipitotemporal infarct.

## Clinical Presentation

- A 45-year-old male without prior medical history presents with right hemiparesis, facial droop, and dysarthria.
- He had a National Institutes of Health (NIH) Stroke Scale of 6 on arrival and tissue plasminogen activator (tPA) was given after 20 minutes.
- After 24 hours, his Glasgow Coma Scale (GCS) decreased to 8 and he was intubated.
- A repeat CT scan demonstrated malignant edema, loss of basal cisterns, and > 5 mm of midline shift (►**Fig. 47.1**).
- Patient was taken for a decompressive craniectomy.
- At follow-up in 6 months he was without speech issues, and ambulating with a mild limp.

## Questions

1. What are the indications and contraindications to administer intravenous (IV) tPA?
2. What are the alternatives to IV tPA? What are their outcomes? Discuss the results of these alternatives.
3. What is the medical management paradigm for this patient?
4. What are the causes of neurological deterioration after stroke? How do you manage cerebral edema in this case?
5. What are the surgical options for this patient? What are the goals of surgery?
6. Discuss surgical considerations of decompressive craniectomy for successful outcomes.
7. Discuss outcomes of decompressive craniectomy.
8. What are the postoperative management considerations for this patient?

## Answers

1. *What are the indications and contraindications to administer intravenous (IV) tPA?*
   - The indications for giving tPA to a patient are:
     - Age 18 years or greater
     - Symptom onset within 3 hours of administration
     - Symptom onset within 3 to 4.5 hours if diagnosis of ischemic stroke is causing measurable neurologic deficit
   - Contraindications for patients treated in the 3- to 4.5-hour window include:
     - Age > 80 years
     - History of both a prior stoke and diabetes
     - Initial NIH Stroke Scale > 25
     - Any oral anticoagulant use regardless of international normalized ratio (INR)[1,2]
   - The absolute contraindications to giving IV tPA to a patient are:
     - Current intracranial hemorrhage (ICH) or subarachnoid hemorrhage
     - History of ICH
     - Intracranial neoplasm, aneurysm, or arteriovenous (AV) malformation
     - Use of therapeutic low-molecular-weight heparin within 24 hours
     - Current use of direct thrombin inhibitors or factor Xa inhibitors
     - Active internal bleeding or acute bleeding diathesis
     - Head trauma, stroke, intracranial surgery, or intraspinal surgery within 3 months
     - Systolic blood pressure ≥ 185 mm Hg or diastolic blood pressure ≥ 110 mm Hg
     - Platelets < 100,000, INR > 1.7, or prothrombin time (PT) > 15
     - Evidence of mutilobar hypodensity on CT scan involving > 33% of the cerebral hemisphere[1,3]
2. *What are the alternatives to IV tPA? What are their outcomes? Discuss the results of these alternatives.*
   - Mechanical thrombectomy
     - The Food and Drug Administration (FDA) approved for recanalization of arterial occlusion in ischemic stroke patients.
     - It can be performed as either a primary therapy or in conjunction with intravascular thrombolysis.
     - Thrombus retrieval systems include those made by Solitaire and Penumbra.
     - Three trials in a meta-analysis—Mechanical Retrieval and Recanalization of Stroke Clots Using Embolectomy (MR RESCUE), Third Interventional Management of Stroke (IMS III), and Multicenter Randomized Clinical Trial of Endovascular Treatment for Acute Ischemic Stroke in the Netherlands (MR CLEAN)—allowed intra-arterial thrombolysis given alone or along with mechanical thrombectomy.
       - MR CLEAN, which also allowed use of intra-arterial urokinase, demonstrated a better functional outcome with a risk ratio (RR) of 1.71 (95% CI, 1.25–2.23),
       - However, all-cause mortality was increased with an RR 1.03 (95% CI, 0.71–1.49) with mechanical thrombectomy.
       - IMS III results showed an improvement in functional outcomes with RR 1.05 (95% CI, 0.86–1.29) and a decreased incidence in all-cause mortality with an RR 0.88 (95% CI, 0.64–1.21).
       - The MR RESCUE trial was the only trial that did not demonstrate better outcomes with mechanical thrombectomy due to incorporation of advanced imaging techniques, which increased time to recanalization.
       - The MR RESCUE trial also stratified patients based on presence or absence of penumbra tissue. Patients with the penumbral pattern demonstrated a worse functional outcome compared to those with nonpenumbral patterns with RR of 0.63 versus 1.00 (95% CI, 0.23–1.72 vs. 0.18–5.46). Both groups did however, demonstrate a decreased incidence in all-cause mortality with penumbral group having an RR of 0.86 (95% CI, 0.32–2.29) and the nonpenumbral group with an RR of 0.67 (95% CI, 0.25–1.78).[4]
     - Overall, according to the results of the meta-analysis including nine studies, mechanical thrombectomy after usual care for large artery occlusion leading to acute ischemic stroke was associated with improved functional outcomes compared to conventional medical treatment

## Answers (*continued*)

alone. There was also no demonstration of increases in ICH as well a reduction trend in all-cause mortality with mechanical thrombectomy compared to usual care alone (IV thrombolysis).[4]

    - Compared to IV-tPA, mechanical thrombectomy was associated with improved functional outcomes as well as higher rates of angiographic revascularization. However, at 90 days, there was not a significant difference in the development of symptomatic ICH or mortality.[1,5]
  - Intra-arterial (IA) fibrinolysis
    - Results of the Prolyse in Acute Cerebral Thromboembolism II (PROACT II) study demonstrated treatment with intra-arterial recombinant prourokinase within 6 hours of acute ischemic stroke onset due to middle cerebral artery (MCA) occlusion had significantly improved outcomes at 90 days (40 vs. 25% with modified Rankin score of 2 or less with $p = 0.04$).
    - As also seen with IV tPA therapy, reduction of time between symptom onset to treatment with IA fibrinolysis correlated with better outcomes.[6]
  - Angioplasty and stenting
    - Enables restoration of anterograde blood flow
    - This is done with or without clot retrieval or fibrinolysis.
    - Studies performed have demonstrated positive results regarding achieving partial or complete recanalization as well as functional outcomes at 1 month post treatment; however, this treatment is not well established and needs further study.[1]

3. *What is the medical management paradigm for this patient?*
   - Current class I evidence for medical management includes treatments to correct any associated tissue hypoxia and hypotension as well as to optimize the patient for eligibility of intravascular thrombolysis. This is achieved by:
     - Giving supplemental oxygen with a goal saturation of > 94%
     - Providing ventilator support in patients with decreased levels of consciousness or other signs of airway compromise
     - Treating sources of hyperthermia with antipyretic agents
     - Cardiac monitoring for at least 24 hours after stroke to assess for the development or arrythmias should also be performed.
     - Blood pressure lowering to < 185 mm Hg systolic and < 110 mm Hg diastolic should be achieved in preparation to administer tPA, followed by blood pressure control for the next 24 hours.
     - IV normal saline may be used to correct hypovolemia and patients should be maintained at normoglycemic levels.
4. *What are the causes of neurological deterioration after stroke? How do you manage cerebral edema in this case?*
   - There are various causes of neurological deterioration after ischemic stroke. They include stroke progression, brain edema, recurrent ischemia, and hemorrhage.
   - Cerebral cytotoxic edema typically peaks at days 3 and 4 post stroke; however, early reperfusion of infarcted tissue has the potential to accelerate the amount of edema within the first 24 hours.
     - This is considered malignant edema.
     - Treatment is geared toward minimizing swelling with hyperosmolar therapy and maintenance of normal intracranial pressures (ICP).
     - When edema leads to increased ICP, the treatment options for lowering ICP include:
       - ○ Mannitol 0.25 to 0.5 g/kg IV
       - ○ Hypertonic saline with sodium goals typically > 145 mEq/L
       - ○ Hyperventilation to $PCO_2$ of 30 to 35 mm Hg
       - ○ Drainage of cerebrospinal fluid (CSF)
       - ○ Surgical decompression[1]
5. *What are the surgical options for this patient? What are the goals of surgery?*
   - Surgical options for this patient include:
     - Decompressive craniectomy (DC)
     - Placement of intraventricular draining catheter
   - The goal of the DC procedure is to remove an adequate area of bone to prevent uncal/transtentorial herniation and relieve pressure on the brainstem.
6. *Discuss surgical considerations of decompressive craniectomy for successful outcomes.*
   - In comparing complications after decompressive craniectomy with or without prior IV tPA administration:
     - Similar rates of new intracranial bleeding or worsening of previous ICH (10% in each group) were observed.
     - The study also demonstrated decreased mortality in patients receiving IV tPA prior to decompressive craniectomy (15 vs. 35%), although the result was not significant with $p = 0.14$. Forty percent of the patients in the study had their DC performed within 24 hours of IV tPA administration and all were without postoperative hemorrhagic complications.[7]
   - When performing an adequate decompressive craniectomy, it is important to assure adequate brain decompression.
     - This is achieved by creating a 12 × 15 cm bone flap and removing the squamous portion of the temporal bone until it is flush with the floor of the middle fossa to assure subtemporal decompression (▶**Fig. 47.2**).

## Answers (*continued*)

  - Smaller-dimension bone flaps, including 6 × 8 cm limited craniotomy flaps were associated with a greater incidence of unfavorable outcomes at 6 months (71.4 vs. 60.2%) compared to the larger craniectomies.[8,9]

7. *Discuss outcomes of decompressive craniectomy*.
   - The Decompressive Surgery for the Treatment of Malignant Infarction of the Middle Cerebral Artery (DESTINY) and Hemicraniectomy and Durotomy on Deterioration from Infarction-Related Swelling Trial (HeADDFIRST) trials established the benefits of hemicraniectomy in reducing mortality in patients with large hemispheric strokes when compared to standard medical management.[10,11]
     - In the HeADDFIRST trial, 21-day mortality was 40% (90% CI [15%, 70%]) in the medical group compared to 21% (90% CI [6%, 47%]) in the surgical group.[10]
     - In the DESTINY trial, survival after 6 and 12 months was 47% in the conservative treatment group versus 82% in the surgical group with a $p = 0.03$ and odds ratio (OR) 5.33 (95% CI, 1.07–26.61).[11]
   - The Hemicraniectomy after Middle Cerebral Artery Infarction with Life-Threatening Edema Trial (HAMLET) did not show a reduction of poor outcomes in patients with hemispheric infarction when treated with surgical decompression within 96 hours of symptom onset with an absolute risk reduction (ARR) of 0% (95% CI, − 21 to 21). However, compared to best medical therapy, surgical decompression lowered case fatality with an ARR of 38% (95% CI, 15–60; $p = 0.002$).[12]
   - A pooled analysis of the results of the DECIMAL, DESTINY, and HAMLET trials demonstrated that decompressive craniectomy performed within 48 hours of stroke reduces mortality (ARR 50%, 34–66) and also increases the number of patients with favorable functional outcomes (ARR 16%, − 0.1 to 33). These trials, however, excluded patients over 60 years of age.[12,13,14]
   - Other studies have also demonstrated benefits when the procedure is performed within 48 hours in patients aged 60 to 80 years. The results showed increased survival without severe disability.[15,16]
8. *What are the postoperative management considerations for this patient?*
   - Postoperatively, patients should have a head CT performed at 12 to 24 hours to evaluate for new hemorrhage or as change in clinical exam or increase in ICP dictates.
   - ICP monitoring with a ventricular catheter or bolt should continue until ICP normalizes.
   - The patient should participate in speech, occupational, and physical therapy as soon as possible.
   - Blood pressure should be managed so as to maximize cerebral perfusion while also avoiding hemorrhage. Recommended systolic pressures < 160 mm Hg for the first 24 hours postoperative.
   - Nursing and supporting staff should be made aware of craniectomy precautions including optimal head positioning to avoid undue pressure on the head.
   - Antibiotic prophylaxis is continued for 24 hours postoperatively or until Jackson Pratt (JP) drain is removed, if placed intraoperatively.[9]

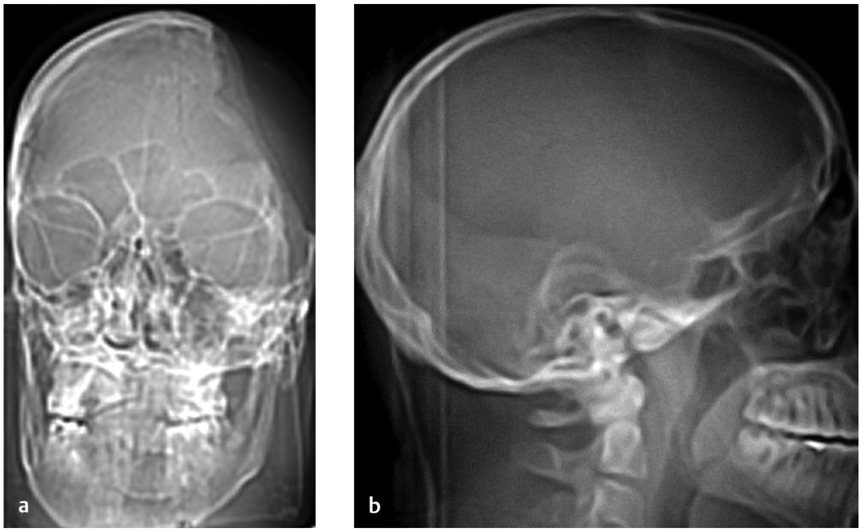

**Fig. 47.2** **(a, b)** Anteroposterior and lateral X-rays post decompression.

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# Case 48 Adult Moyamoya

*Anthony M. T. Chau, Cristian Gragnaniello, Remi Nader, Anthony J. Caputy, Dimitri Sigounas, and Marguerite Harding*

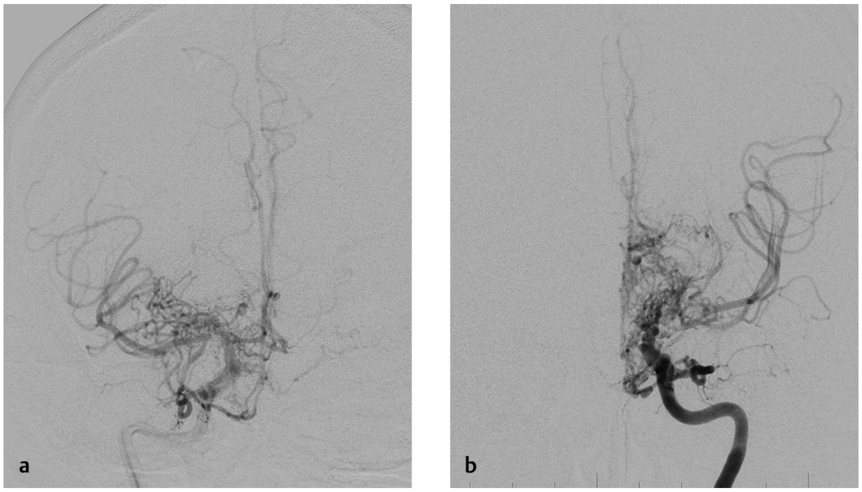

**Fig. 48.1** (**a**) Digital subtraction angiography image from right internal carotid artery (ICA) injection in the anteroposterior plane. (**b**) Left ICA injection in the same patient.

## Clinical Presentation

- A 46-year-old Caucasian woman presents following the sudden onset of transient dysarthria and mild left-sided facial droop.
- The symptoms spontaneously resolved within a few hours.
- A noncontrast CT brain was unremarkable for hemorrhage or infarct.
- MRI brain demonstrated evidence of small vessel ischemic changes in the watershed region between the anterior cerebral artery (ACA) and middle cerebral artery (MCA) territories on the right, without evidence of acute infarct.
- Abnormalities of the anterior circulation bilaterally were identified.
- A 6-vessel angiogram was performed.

## Questions

1. Describe the imaging findings.
2. What is Moyamoya disease (MMD)?
3. What are the pathophysiological mechanisms of MMD?
4. What imaging modalities are available to better evaluate and study patients with MMD?
5. What is the epidemiology of adult MMD?
6. What is the natural history of adult MMD? What have observation studies shown?
7. What are the treatment options?
8. Describe briefly the principles of an encephaloduroarteriosynangiosis (EDAS) procedure.

## Answers

1. *Describe the imaging findings.*
   Digital subtraction angiography (DSA) demonstrates bilateral stenosis of the terminal internal carotid arteries (ICAs) and proximal ACAs and MCAs. There are bilateral basal lenticulostriate networks of fine collateral vessels.
2. *What is Moyamoya disease (MMD)?*
   - Idiopathic bilateral steno-occlusive disease of the terminal ICAs and proximal ACAs and MCAs, with characteristic basal lenticulostriate “moyamoya” collateral vessels.
   - Suzuki (1983) described six angiographic stages:[1]
     - Stage 1: carotid fork narrowing
     - Stage 2: initiation of basal moyamoya
     - Stage 3: intensification of moyamoya, with involvement of the ACAs and MCAs
     - Stage 4: minimization of moyamoya, with involvement of the posterior cerebral artery

## Answers (*continued*)

  - Stage 5: reduction of moyamoya, with absence of all the main cerebral arteries
  - Stage 6: disappearance of moyamoya, with cerebral blood flow (CBF) supplied through the external carotid system
- Atypical MMD:[2]
  - Noncarotid steno-occlusive vasculopathy
  - Associated aneurysms or arteriovenous malformations
- Moyamoya syndrome:[3]
  - When the characteristic vasculopathy is only unilateral. Note, however, that MMD can occur with asymmetric severity, and that 30 to 40% of patients will develop contralateral disease at 5 years.
  - When the characteristic angiographic features occur in patients with recognized risk factors, such as Down's syndrome, neurofibromatosis type 1, sickle cell disease, post therapeutic cranial irradiation, etc.

3. *What are the pathophysiological mechanisms of MMD?*
   - On histopathological analysis, main vessel occlusion occurs from smooth muscle hyperplasia and luminal thrombosis, rather than from atheromatous or inflammatory changes.[3,4] The intima is eccentrically thickened and the internal elastic lamina is tortuous or duplicated.
     - The pathophysiology is speculated to involve primary vessel wall injury followed by abnormal wall remodeling.
     - The process is perhaps mediated by aberrant immunological targeting and caspase-dependent apoptosis.
     - Bone marrow-derived vascular progenitor cells, rather than adjacent smooth muscle cells from the tunica media, may aggregate on diseased vessels and differentiate into the pathological neointimal smooth muscle cells.[4]
   - Moyamoya collaterals are either preexisting or newly recruited dilated perforating arteries with fragmented elastic lamina and thinned media.
   - Some may harbor microaneurysms, while others may be collapsed and thrombosed.
   - Genetic predisposition is suspected, especially given the high incidence among patients of Asian origin.
     - Ten percent of Japanese and 6% of American MMD patients have a first-degree relative also having the condition.
     - An autosomal dominant pattern with incomplete penetrance has been proposed.
     - Possible genetic associations from genome-wide linkage studies of Japanese families affected with MMD include loci on chromosomes 3, 6, 8, 12, and 17; 17q25 has been especially identified as a candidate locus.[5]
     - There are case reports of identical twins with only one affected sibling, suggesting an environmental trigger may also be involved.
4. *What imaging modalities are available to better evaluate and study patients with MMD?*
   - MRI
     - Conventional T2-weighted imaging may demonstrate loss of usual vascular flow voids of the major anterior circulation arteries, accompanied by numerous flow voids of the basal cisterns and basal ganglia.
     - Diffusion-weighted imaging (DWI) or fluid-attenuated inversion recovery (FLAIR) may demonstrate evidence of new or old ischemic injury, respectively. Watershed infarcts are common. Asymptomatic microhemorrhages may be present in the periventricular white matter.
     - FLAIR or gadolinium-enhanced T1-weighted imaging may demonstrate the so-called ivy sign—leptomeningeal enhancement. This is a characteristic albeit nonspecific sign of MMD, representing leptomeningeal flow engorgement.
     - Adequate MR angiography (MRA) may demonstrate steno-occlusive vessels, basal cerebral moyamoya vessels, and transdural collaterals. MRA can be an acceptable modality for the diagnosis of MMD (i.e., formal angiography is not always necessary).
   - Metabolic studies provide information on the tissue metabolism of the brain (direct) or cerebrovascular autoregulatory function (an indirect measure).
     - Metabolic demand is assessed noninvasively through measurement of the oxygen extraction fraction (OEF).
     - Cerebrovascular reactivity (CVR) is assessed before and after hypercapnic challenge, either through intravenous acetazolamide (administration of this carbonic anhydrase inhibitor produces a shift toward carbonic acidosis), or end-tidal $CO_2$ manipulation through a rebreathing device. The response in normal subjects is an increase in CBF. In MMD patients with perpetually dilated arterioles and hence exhausted cerebrovascular reserve, no change or a reduction (steal phenomenon) compared to baseline is observed.
   - CBF is measured by observing an introduced tracer over a period of time.
     - The mean transit time (MTT) and cerebral blood volume (CBV) can be measured according to the tracer concentration in a region of interest over a period of time.[2]
     - CBF is measured using the central volume theorem, i.e., CBF = CBV/MTT. Decreased CBF in MMD is thought to correlate with parent vessel occlusion.

## Answers (*continued*)

- Positron emission tomography (PET) requires an onsite cyclotron or linear accelerator to generate its short half-life radiotracers.
    - PET measures OEF to assess the metabolic demand of the brain. Radiolabelled $^{15}O_2$ is injected intravenously and the proportion of oxygen extracted from the blood is measured. The OEF is increased in ischemic brain.
    - PET can also measure CBF directly using $H_2{}^{15}O$ and CBV using $C^{15}O$.
- Xenon-enhanced CT measures the autoregulatory response after hypercapnic/acetazolamide challenge. The radio-opaque gas is inhaled and diffuses throughout tissue according to the blood flow. It is associated with various side effects such as vomiting, headaches, convulsions, and respiratory failure.
- Perfusion CT is a rapid means of assessing CBF using iodinated contrast, but is susceptible to artifacts relating to blood pressure, hematocrit, and motion. The flow input to the pathological tissue is required to calculate CBF. The choice of a reference artery for this flow calculation is particularly problematic in MMD patients with bilateral steno-occlusive disease. The posterior cerebral artery may be an option. Perfusion MRI is similar to its CT counterpart, but uses gadolinium. Acetazolamide challenge can be employed.
- Single-photon emission CT (SPECT) assesses CVR using a gamma-ray-emitting tracer, such as $^{123}$I, N-isopropyl-p-iodoamphetamine autoradiography ($^{123}$I-IMP-ARG) or technetium hexamethyl propylene amine oxide (99-m Tc-HMPAO), which is injected intravenously (▶**Fig. 48.1**).[4] The tracer crosses the blood–brain barrier and is temporarily fixed to brain tissue, where it is measured before and after acetazolamide challenge. Typically, this is performed over a 2-day period to allow for tracer washout (see ▶**Fig. 48.2** for example of SPECT study).
- Blood oxygen level-dependent (BOLD) MRI assesses CVR through differences in the paramagnetic signal of deoxygenated hemoglobin (deoxyHb) and oxygenated hemoglobin (oxyHb), detected on heavily weighted T2 sequences. After hypercapnic challenge, the action of local capillaries leads to a drop in oxyHb, and an increase in deoxyHb and $CO_2$. Following a 2- to 6-second lag, a rebound surge of oxyHb occurs, washing away deoxyHb. Compared to nuclear medicine techniques, BOLD MRI confers advantages with its high spatial and temporal resolution, no requirement for radiopharmaceuticals, and challenge testing can be obtained at the same sitting. In addition, no arterial input function is required.

5. *What is the epidemiology of adult MMD?*
    - In Japan, the annual incidence of MMD ranges from 0.35 to 0.94 per 100,000, with a prevalence from 3.16 to 10.5 per 100,000.[6,7] A bimodal distribution is observed with peaks in children aged 5 to 14 years and adults in their fifth decades of life. There is a two-fold higher incidence in women for sporadic cases, and 5:1 ratio in familial cases.[8]
    - In the United states, the incidence is lower at 0.086 per 100,000, but with a higher incidence of 0.28 in the US Asian subpopulation.[9] A bimodal distribution is also observed, at ages 5 to 9 years and 55 to 59 years. The female-to-male ratio is similarly 2:1 in US and European populations.[10]
    - The phenotype of MMD varies between Asian and Caucasian patients. The most frequent presentation, particularly in Caucasian adults, is an ischemic event (more than two-thirds of patients). Patients of Asian origin have a higher predisposition than Caucasians to present with intracranial hemorrhage (approximately a quarter of patients).[7,11,12]
6. *What is the natural history of adult MMD? What have observation studies shown?*
    - Adult MMD may present with ischemia, hemorrhage, seizures, incidentally or other (such as headaches).
    - Adult MMD is considered a progressive disease regardless of presentation.
    - The course may be latent, slowly indolent, intermittent, or rapid.[13]
    - Kuroda (2005) followed 52 Japanese adults heterogeneously diagnosed with typical MMD for a mean period of 6 years.[14] These patients were a subset of 120 subjects not chosen for surgical intervention.
        - Thirteen percent patients experienced an ischemic event or worsening of cerebral hemodynamics.
        - Twenty-one percent patients demonstrated progressive occlusive disease on subsequent imaging (27% of whom developed posterior circulation involvement).
        - Female gender was a significant predictor of disease progression.
    - Kuroda (2007) also examined 40 patients with asymptomatic MMD with a mean follow-up period of 3.5 years.[15]
        - Twenty percent of patients had evidence of ischemic infarction or disturbed cerebral hemodynamics at recruitment.
        - Advanced age was associated with more severe angiographic staging.
        - Six patients underwent surgical revascularization.
        - Of the remaining 34, 9% experienced transient ischemic attack (TIA), 3% infarct, and 9% hemorrhage during the follow-up period.
        - The annual risk for any stroke was 3.2%.
    - In a North American population, Hallemeier (2006) observed a 5-year stroke rate of 82% in 17 MMD patients presenting with infarction.[16]
    - Kraemer (2008) similarly reported that 81% of 21 European MMD patients suffered strokes after

## Answers (*continued*)

their first ischemic event, most within the first 2 years.[17]

- Kobayashi (2000) found that one-third of 42 conservatively managed hemorrhagic MMD patients rebled at a rate of 7% per patient per year.[18]
  - Rebleeding location suggests a diffuse vulnerability of the periventricular collateral vessels.
  - Rebleeds significantly affected the rate of a good recovery (46 vs. 21%) and mortality (7 vs. 29%), and in 36% of cases rebleeding occurred more than 10 years afterwards.
- Similarly, in a study of 36 hemorrhagic MMD patients followed for a mean of 13 years, Morioka (2003) found a high rate of rebleeding (61%) which was associated with an unsatisfactory clinical outcome.[19]
- Festa (2010) found that two-thirds of adults with MMD demonstrated neurocognitive dysfunction, including impairment in processing speed, verbal memory, verbal fluency, and executive function, likely related to frontotemporal perfusion impairment.[20] Twenty-eight percent patients reported moderate to severe depression.

7. *What are the treatment options?*
   - Surgical revascularization improves the natural history of MMD.[21]
   - Indirect, direct superficial temporal artery to middle cerebral artery (STA-MCA), and combined bypass procedures are possible.
   - Indirect procedures include EDAS, or pial synangiosis, encephalomyosynangiosis (EMS), and dural inversion.
     - Indirect bypass has the benefit of shorter operative time and fewer technical demands compared to direct bypass.
     - However, the natural history is not altered until sufficient collateral supply develops over several months.
     - Unlike pediatric patients, collateral anastomoses following indirect bypass are known to develop poorly in adults, perhaps in 50% of patients.[22]
   - However, the long-term postoperative stroke rates in adult MMD is similar irrespective of the above procedure chosen:
     - Indirect bypass: 0 to 11%[23]
     - Direct bypass:
       - Five-year stroke rate 5.5%[11,21]
       - In cases of hemorrhagic MMD, a 4% rebleed/mortality risk was observed at 2 years.[24]
     - Combined bypass: 5 to 13%[23]
   - Institutional preference for indirect or direct bypass may dictate the type of surgical revascularization performed, as both forms currently appear equally effective in preventing stroke.[25]
8. *Describe briefly the principles of an encephaloduroarteriosynangiosis (EDAS) procedure.*
   The steps of an EDAS procedure are as follows:[2]
   - After preparing the scalp, the STAs and occipital arteries (OcAs) are outlined using a Doppler ultrasound; then the skin overlying them is opened and the vessels are identified.
   - The STAs and OcAs are separated from neighboring tissues along with a strip of adventitia on either side of the vessels.
   - A craniotomy or a small craniectomy is performed, and then a Z-shape durotomy is tailored.
   - The adventitia is sutured to close the durotomy edges, making the STAs and the OcAs in close proximity to pial vessels.
   - This facilitates collateral development over time, from the external to the intracranial circulation.

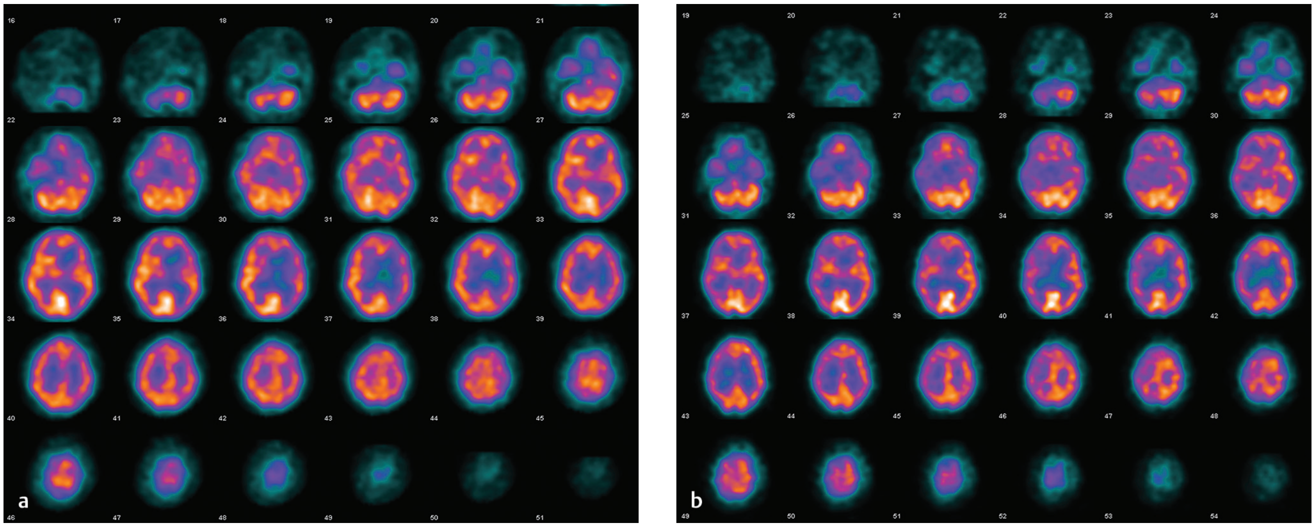

**Fig. 48.2** Single-photon emission CT (**a**) pre- and (**b**) post-acetazolamide challenge, demonstrates mildly reduced cortical perfusion in the right frontotempoparietal lobes in keeping with abnormal cerebrovascular reserve.

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# Case 49 Hypertensive Putaminal Hematoma

*Nicholas J. Erickson, Cristian Gragnaniello, and Remi Nader*

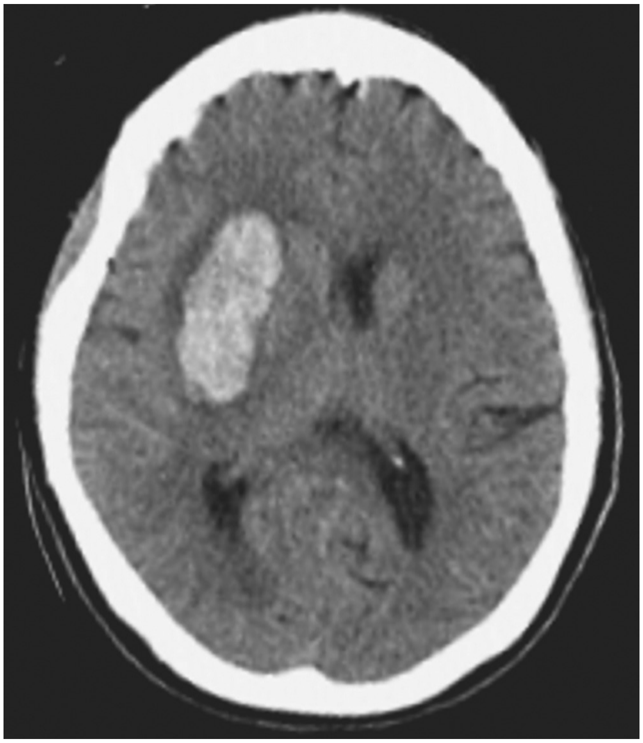

**Fig. 49.1** Follow-up CT scan without contrast at the level of the basal ganglia.

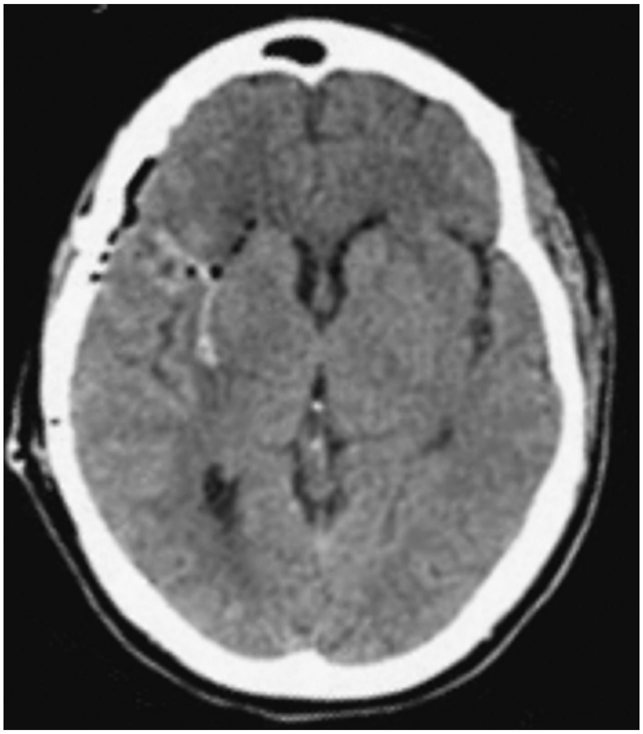

**Fig. 49.2** Postoperative CT scan without contrast at the same level of the basal ganglia.

## Clinical Presentation

- A 56-year-old woman with diabetes mellitus, hypertension, and obesity presents with sudden-onset left-sided weakness.
- Initial studies show a 2-cm right-sided basal ganglia intracerebral hematoma (ICH).
- She is managed conservatively in the intensive care unit (ICU) initially; however, she starts becoming lethargic on day 5 and her weakness is more pronounced.
- A follow-up CT scan is obtained (▶**Fig. 49.1**).

## Questions

1. What are the criteria to operate on a basal ganglia ICH?
2. What would you recommend in this case?
3. What are the surgical and nonsurgical options?
4. What are the expected outcomes based on each option?

   You decide to evacuate the ICH with a craniotomy. The procedure goes well, but postoperatively, the patient is still lethargic and hard to arouse; she remains this way for over 12 hours. A postoperative CT scan is obtained (▶**Fig. 49.2**).
5. Provide a differential diagnosis.
6. What is your management now?

## Answers

1. *What are the criteria to operate on a basal ganglia ICH?*
   - The recent International Surgical Trial in Intracerebral Haemorrhage (STICH) trial[1] suggests that prophylactic removal of ICH had no clear advantage over medical management with removal only if deterioration occurs.[1]
   - Some of the reasons to favor surgery have been:[1–5]
     - Symptomatic patients with potential for acceptable recovery who seem to be worsening rapidly despite medical management
     - Moderate ICH volume (~10–30 mL)
     - Favorable location
     - Younger age
     - Edema
     - Midline shift
2. *What would you recommend in this case?*
   - This patient does fit some of the current criteria:
     - She is relatively young (56 years old).
     - The location is favorable (right-sided, nondominant).
     - The size is favorable (~ 20–30 mL).
     - There is midline shift as well as some mass effect.
     - There has been a deterioration in her mental status.
   - Given these factors, surgery should be offered as an option to the family with clear understanding that expected outcome may only be slightly improved at best. Also, observation is a reasonable option as well, given the family's wishes (▸**Fig. 49.3**). In this case, the family did opt for surgery.
3. *What are the surgical and nonsurgical options?*
   - Small craniotomy and resection
   - Stereotactic technique for drainage of hematoma[5]
   - Endoscopic removal of ICH[3]
   - Placement of catheter in the ICH ± tissue plasminogen activator (tPA) infusion
   - Ultrasound (Cavitron Ultrasonic Surgical Aspirator [CUSA]) aspiration
4. *What are the expected outcomes based on each option?*
   - There is no significant difference in long-term outcomes between observation and surgery in general regarding deep-seated brain ICH.
   - Overall favorable outcome is 25%.
   - The absolute difference in favorable outcome between surgery and observation (based on STICH study) is 2 to 3%.[1]
   - Minimally Invasive Surgery plus rt-PA for Intracerebral Hemorrhage Evacuation (MISTIE) is a series of clinical trials that demonstrated that minimally invasive techniques along with intraclot rtPA administration followed by a sterile flush reduced the rates of death and improved neurologic function and quality of life in the year following the ICH.
   - MISTIE II trial demonstrated significant reduction in perihematomal edema in the treatment group (minimally invasive surgery [MIS] + rtPA) and a trend toward improved outcomes. A phase 3 trial is currently in progress.[6]
5. *Provide a differential diagnosis.*
   - Swelling/edema
   - Infarction/stroke
   - Seizures
   - Metabolic deterioration (hypo/hypernatremia, hypoxia, hypercapnia)
   - Infection
   - Side effect of medications
   - Endocrinopathy
6. *What is your management now?*
   - Given the fact that the postoperative CT (▸**Fig. 49.2**) shows complete resolution of the hematoma with no significant mass effect, midline shift, or other evidence of increased intracranial pressure (ICP), the problem remains nonsurgical at this point.
   - Medical management in ICU
     - Laboratory work-up: complete blood count, electrolytes, coagulation profile, drug levels, liver function tests, and endocrine panel
     - Cultures: blood, urine, sputum, and cerebrospinal fluid (lumbar puncture)
     - Seizure prophylaxis medications
     - Intravenous fluids
   - May consider MRI if infarct is suspected

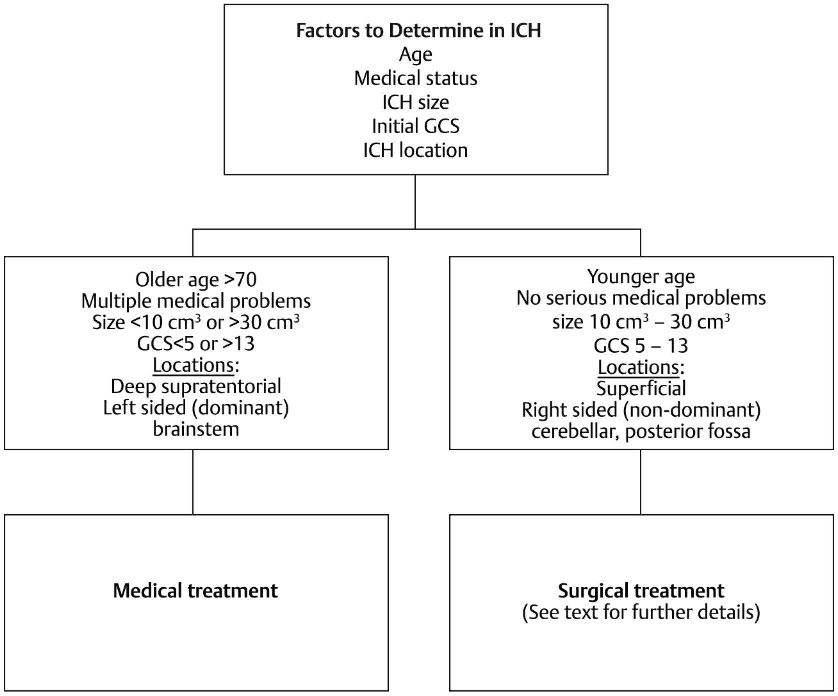
Factors to Determine in ICH
Age
Medical status
ICH size
Initial GCS
ICH location
Older age >70
Multiple medical problems
Size <10 cm³ or >30 cm³
GCS<5 or >13
Locations:
Deep supratentorial
Left sided (dominant)
brainstem
Younger age
No serious medical problems
size 10 cm³ – 30 cm³
GCS 5 – 13
Locations:
Superficial
Right sided (non-dominant)
cerebellar, posterior fossa
Medical treatment
Surgical treatment
(See text for further details)

**Fig. 49.3** Guidelines for the treatment of intracerebral hemorrhage. GCS, Glasgow Coma Scale; ICH, intracerebral hemorrhage. (Adapted from Mendelow et al 2005; Broderick et al 1999)[1,4]

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# Case 50 Cerebellar Hemorrhage

*Julius July and Eka Julianta Wahjoepramono*

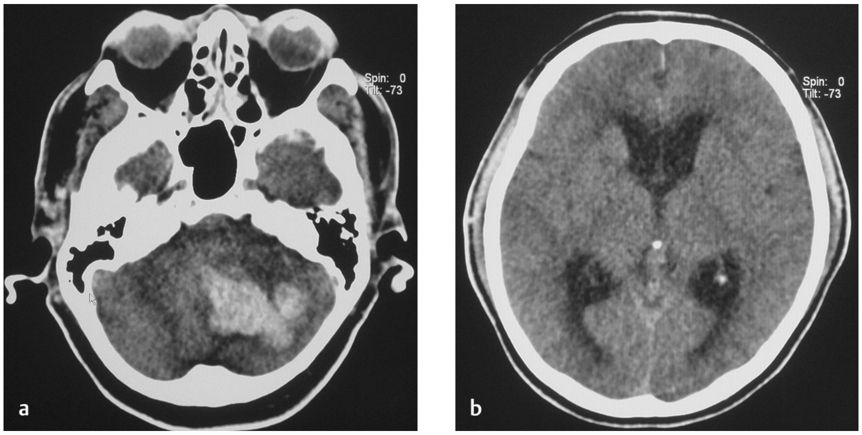

**Fig. 50.1** Noncontrast enhanced CT scan showing left cerebellar hemorrhage (**a**) with effacement of fourth ventricle and (**b**) enlargement of the lateral ventricles.

## Clinical Presentation

- A 55-year-old man with a long history of poorly controlled hypertension, diabetes mellitus, and history of heavy smoking presents to the emergency room with acute decreased level of consciousness.
- This morning, he woke up with a headache that has progressively gotten worse.
- On physical examination, he opens his eyes spontaneously and localizes the pain. There is no obvious weakness in his extremities. Pupils are both 3 mm and responsive to light. Initial blood pressure is 200/110 mm Hg.
- A CT scan is obtained and shown in ▶**Fig. 50.1**.

## Questions

1. Describe a differential diagnosis.
2. What is the most likely diagnosis?
3. Discuss recommendation for surgical intervention on cerebellar hemorrhage.
4. Describe your surgical plan.
5. After you open the dura, you observe a very swollen and tight cerebellum. What are your steps in managing intraoperative brain swelling?
6. Describe your postoperative care.

## Answers

1. *Describe a differential diagnosis.*
   - Hypertensive bleeds account for most cerebellar hemorrhages.
   - A small number of patients can present with a small aneurysm of the posterior circulation that, if ruptured, can give a similar clinical picture.[1]
   - The frequency of cerebellar hemorrhage ranges between 5 and 10% of all intracranial hemorrhages (ICHs), and it occurs predominantly in the older age groups in their sixth to the eighth decades of life.
   - Cerebellar cavernoma bleeding should be considered if the clot is well circumscribed, and usually the symptoms are relatively mild compared to the size of the hematoma (▶**Fig. 50.2**) and require treatment very rarely.
   - A posterior fossa arteriovenous malformation (AVM) with prenidal aneurysm should always be considered and needs to be ruled out from the differential diagnosis. This lesion should be treated aggressively because it has a significantly higher risk for rupture, almost 9 times higher than supratentorial AVM (41 vs. 4.7%).[2]
   - The history of diabetes in this case is also considered a general risk factor for developing a cerebellar hemorrhage.
   - Intratumoral bleeding should also be considered in the differential diagnosis. Cerebellum is a common location for brain metastases, and some tumor may be prone to bleed. Usually the CT scan will show a heterogenous density appearance with significant surrounding edema.
2. *What is the most likely diagnosis?*
   - The cerebellar hemorrhage seen on CT scan is most likely due to hypertension.
3. *Discuss recommendation for surgical intervention on cerebellar hemorrhage.*
   - Recommendation for surgical intervention include:
     – Patients with Glasgow Coma Scale (GCS) score ≤ 13 or with hematoma size ≥ 4 cm
     – Ventricular catheter placement is recommended for patients with hydrocephalus and no coagulopathy. Most cases with hydrocephalus also require evacuation of the hematoma.
     – The presence of the “tight posterior fossa” (TPF) concept, described by Weisberg et al,[3] warrants surgical intervention. Weisberg defines this as:
       ○ Obliteration of the basal cisterns of the posterior cranial fossa
       ○ Enlargement of the third ventricle, lateral ventricles, and temporal horns
       ○ Effacement of the fourth ventricle
     – The TPF does not only depend on the size of the hematoma. A hematoma of similar size may exert widely different amounts of compression influenced by several factors such as patient’s age, the amount of cerebellar atrophy, and the anatomy of the posterior fossa. Based on the TPF concept, the critical size for hematoma evacuation can be reduced to 5–10 mm from 3 cm.[1,3]
4. *Describe your surgical plan.*
   - Initially, a ventricular catheter is inserted (one may choose the entry site at Kocher’s point or at Frazier burr hole site). Two to 3 cc of cerebrospinal fluid (CSF) is drained slowly to control the intracranial pressure (ICP), then the drainage is stopped to avoid upward herniation.
   - A suboccipital craniectomy is then performed with goal of removing the ICH, decompressing the brainstem, and relieving the hydrocephalus. The external drain can be opened after dural opening.
   - A transverse incision along the folia, closest to the most superficial part of the clot is performed. After removing the ICH, meticulous hemostasis should be obtained. The blood clot should be sent to pathology for examination.
   - Very often, the cerebellum itself will swell after the surgery. It is better to close the dura via a duraplasty, using pericranium, fascia lata, or synthetic dura.
   - The ventricular catheter in general is kept temporarily. However, some cases necessitate conversion to a permanent ventriculoperitoneal shunt.[1,4]
5. *After you open the dura, you observe a very swollen and tight cerebellum. What are your steps in managing intraoperative brain swelling?*
   - The following is a checklist of steps to ensure proper management of intraoperative posterior fossa swelling:[5,6]
     – Elevate the patient’s head to above 30 degrees.
     – Check neck positioning for obstruction of venous return and readjust as necessary.
     – Hyperventilate to a $PCO_2$ of 30 to 35 mm Hg.
     – Infuse a dose of mannitol and/or Lasix (Aventis Pharmaceuticals, Parsippany, NJ).
     – Ensure adequate sedation and pharmacologic muscle paralysis.
     – Decompress the hematoma rapidly.
     – Drain some CSF by ventriculostomy.
     – Open the cisterna magna and drain some more CSF.
     – One may need to resect parts of the cerebellar hemisphere.
     – Ensure that the foramen magnum is open.
     – If the swelling is still uncontrollable, then consider the following causes:
       ○ Intraparenchymal hematoma in a different location
       ○ Contralateral or supratentorial subdural or epidural hematoma
       ○ Cytotoxic edema from trauma
       ○ Venous infarction
     – In intractable cases, some of the following steps may be considered:
       ○ Duraplasty and quick closure

## Answers (*continued*)

    - Leaving the bone flap out (craniectomy)
    - Obtaining an emergent CT scan of the head and considering going back to the operating room for further exploration.
    - In the past, exploratory supratentorial burr holes were suggested. However, with the advent of fast spiral CT scanners and/or mobile intraoperative CT scanners, an adequate scan can be completed within minutes eliminating the need and potential risks associated with such a procedure.

6. *Describe your postoperative care.*
    - Postoperatively, the patient should be monitored in the intensive care unit (ICU) for at least 24 hours in anticipation of the peak of swelling. Blood pressure should be closely monitored with an arterial line and controlled closely with intravenous (IV) antihypertensives (e.g., nitroprusside or nicardipine).
    - Patients with a critical neurologic condition remain under mechanical ventilation and in certain cases, consideration of corticosteroids may be reasonable (dexamethasone 10 mg IV once, then 6 mg IV every 6 hours).
    - If required, other agents may be administered to reduce the ICP.
    - A postoperative CT scan should be obtained within 24 hours (▶**Fig. 50.3**).
    - As the patient's neurologic condition improves, he may be transferred to the neurosurgical ward and subsequently discharged from the hospital once the blood pressure remains under control with oral antihypertensives.[1,4]

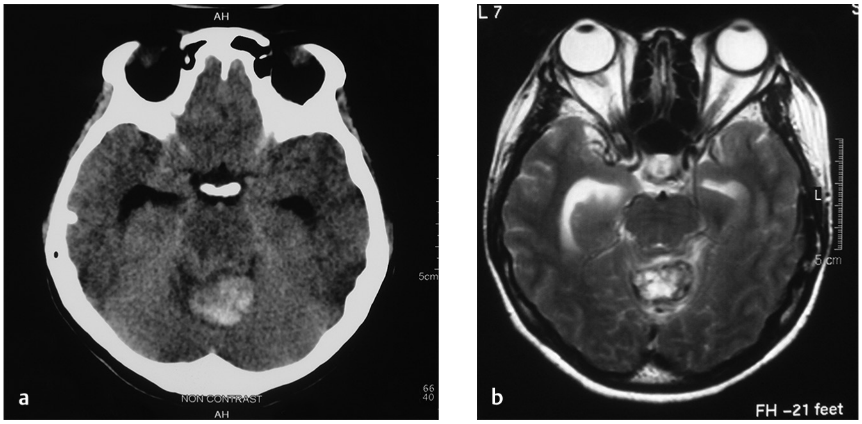

**Fig. 50.2** Noncontrast CT scan (**a**) shows a midline cerebellar hemorrhage, well demarcated, compressing the posterior part of the brainstem and the aqueduct, causing the temporal horn dilatation. The T2-weighted MRI (**b**) shows heterogenous intensity (popcorn-like appearance) of early and late subacute blood. It is consistent with cavernoma bleeding.

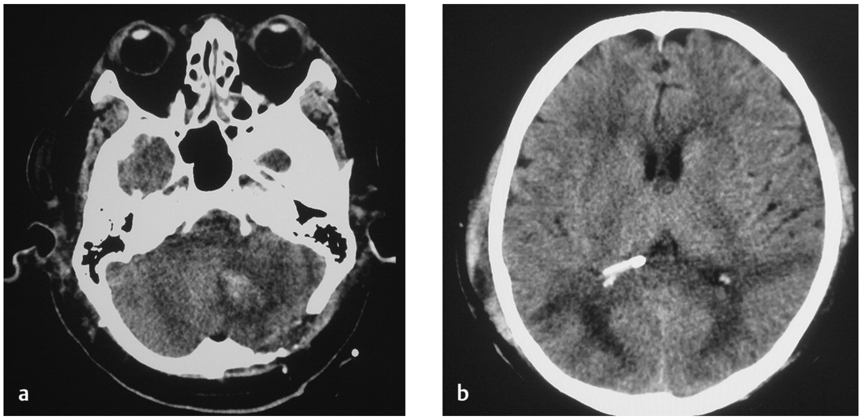

**Fig. 50.3** (**a**) Postoperative CT scan shows that most of the blood clot has been removed from the left cerebellum, but the posterior fossa is still very tight. (**b**) The CT scan confirms the ventricular catheter placement, and the reduction in size of the lateral ventricles.

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# Case 51 Amaurosis Fugax with Carotid Occlusion

*Glenn C. Hunter, Alwin Camacho, and Craig C. Weinkauf*

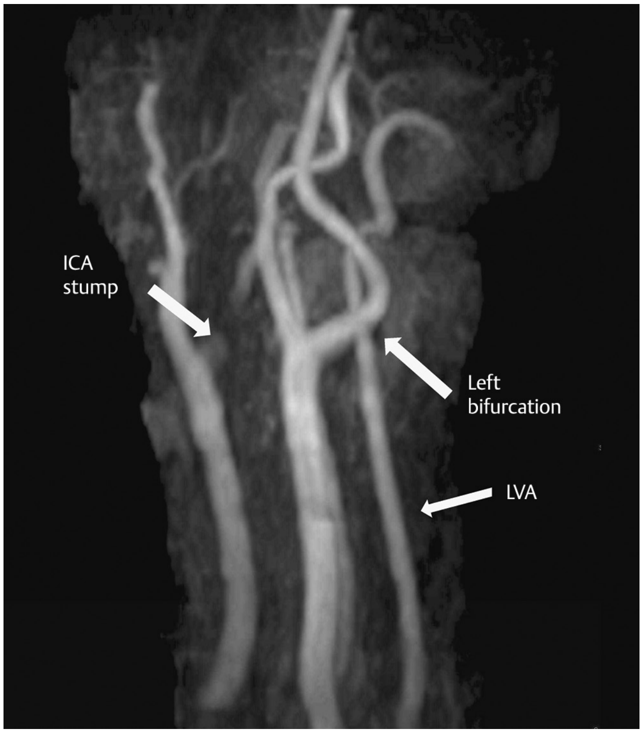

**Fig. 51.1** MR angiography of the neck showing a right internal carotid artery (ICA) stump, the patent left carotid bifurcation, and left vertebral artery (LVA).

## Clinical Presentation

- A 70-year-old man presents to his ophthalmologist with worsening vision in the right eye.
- Three months previously he experienced an episode of amaurosis fugax.
- A funduscopic exam shows a Hollenhorst plaque and diminished flow in a branch of the retinal artery.
- He is referred for urgent neurosurgical consultation.

## Questions

1. List the differential diagnoses.
2. What are the most common causes of monocular visual symptoms?
3. What are the most common sources of thromboembolism to the eye?
4. Physical examination reveals bilateral carotid bruits and a blood pressure of 145/95 mm Hg. What studies would you obtain?
   A carotid duplex scan and MR angiography (MRA) are ordered. The carotid duplex shows right internal carotid artery (RICA) occlusion with < 50% stenosis on the left. MRA shows RICA occlusion with a patent external carotid artery (ECA; ▶**Fig. 51.1**). The left carotid artery appears normal.
5. Outline your plan of management.
6. During your evaluation, he experiences another episode of worsening vision. What are the potential causes of this episode?
7. How may this episode alter your treatment plan?
8. In view of the results of the MRA, would you order a cerebral angiogram?
9. A four-vessel angiogram is obtained (▶**Fig. 51.2**). Interpret the findings on the right.

## Questions (*continued*)

10. What is the significance and importance of a residual carotid stump?
11. What are your therapeutic options?
12. You consider doing a common carotid endarterectomy and excision of the stump. What is the risk and natural history of stroke in patients who present with monocular visual symptoms versus those who present with transient ischemic attacks (TIAs)?
13. What are the risk factors for stroke in patients with monocular visual symptoms?
14. How would you treat this patient?

## Answers

1. *List the differential diagnoses.*
   - Transient monocular blindness and amaurosis fugax are umbrella terms describing a range of patterns of transient monocular visual field loss. The incidence rises from ≈1.5/100,000 in the third decade of life to ≈32/100,000 in the seventh decade of life.[1]
   - Ocular manifestations account for 15% of TIAs. The differential diagnoses of this patient's symptoms include:[1–5]
     – Amaurosis fugax
     – Hollenhorst plaques
     – Retinal artery occlusion (central/branch)
     – Ocular ischemic syndrome
     – Venous stasis retinopathy
     – Nonspecific visual symptoms
     – Autoimmune diseases (systemic lupus erythematosus, scleroderma rheumatoid arthritis, and antiphospholipid syndrome)
     – Familial hemiplegic migraine
     – Giant cell arteritis
     – Carotid dissection
2. *What are the most common causes of monocular visual symptoms?*
   - Visual symptoms in patients in this age group are mostly due to thromboembolism and less often due to hypoperfusion.[6,7]
3. *What are the most common sources of thromboembolism to the eye?*
   - The carotid bifurcation is the most common embolic source in these patients.[6]
   - Cardiac thromboembolism (atrial fibrillation, patent foramen ovale, aortic valvular disease, and aortic arch atheroma), intravascular injection of talc, and steroidal suspensions are other potential sources.
4. *Physical examination reveals bilateral carotid bruits and a blood pressure of 145/95 mm Hg. What studies would you obtain?*
   - Although the focus of the studies should be to find an embolic source, autoimmune etiologies also need to be excluded. A carotid duplex scan should be the initial screening test.[2,7]
5. *Outline your plan of management.*
   - Admission to the hospital
   - Locate potential cerebral embolic infarcts and exclude intracranial hemorrhage with CT and/or MRI and embolic sources with CT angiography (CTA) and MRA.
   - Exclude cardiac embolic sources by obtaining electrocardiogram (EKG), echocardiogram, and transesophageal echocardiogram.
   - Specific laboratory tests: complete blood count (CBC), erythrocyte sedimentation rate, C-reactive protein, serum electrolytes, blood glucose, coagulation profile, and autoimmune panel
   - Antiplatelet therapy
6. *During your evaluation, he experiences another episode of worsening vision. What are the potential causes of this episode?*
   - Worsening symptoms may be due to hypoperfusion or another embolic episode. Embolic events are usually more common than hypoperfusion.[2,5]
7. *How may this episode alter your treatment plan?* In patients with carotid occlusion, control of blood pressure to maintain cerebral perfusion is an important consideration. Also, if the patient is not on antiplatelet therapy or is only on aspirin, an additional agent, such as clopidogrel, should be added or the patient should be commenced on a combination of aspirin and dipyridamole (Aggrenox). Heparin should be considered if the ipsilateral occlusion is recent and there is no evidence of intracranial hemorrhage.[4]
8. *In view of the results of the MRA, would you order a cerebral angiogram?*
   - The presence of persistent symptoms may warrant the following studies: a four-vessel cerebral angiogram allows assessment of the ECA and ophthalmic arteries and their collateral communication, and it will also eliminate a "string sign." A digital subtraction angiography (DSA) is both diagnostic and interventional. However, it is an invasive procedure, as it uses ionizing radiation, carries itself a risk of embolic strokes, and is not ideal for multiple follow-up exams.
   - MRA may be done instead of a DSA; it has a good negative predictive value for excluding 50 to 99%

## Answers (*continued*)

stenosis but has a low positive predictive value. MRA may miss a string sign or overread the degree of stenosis.
- CTA can be done faster, is widely available, and may be a better study in unstable patients, but it may be limited by calcification, ossification, and renal impairment.

9. *A four-vessel angiogram is obtained (*▶**Fig. 51.2***). Interpret the findings on the right.*
   - The four-vessel angiogram shows occlusion of the RICA at its origin, with a residual stump and antegrade vertebral flow.
10. *What is the significance and importance of a residual carotid stump?*
    - The residual stump may be an embolic source and if associated with a high-grade ECA stenosis, may contribute to retinal hypoperfusion.[3,6,7]
11. *What are your therapeutic options?*
    - The therapeutic options include doing a common carotid endarterectomy and excision of the stump or placing a common carotid stent and occluding any residual lumen of the stump.
12. *You consider doing a common carotid endarterectomy and excision of the stump. What is the risk and natural history of stroke in patients who present with monocular visual symptoms versus those who present with transient ischemic attacks (TIAs)?*
    - The 3-year risk of ipsilateral stroke is 10% for patients with transient monocular blindness versus 20% for those presenting with hemispheric symptoms.[8–12]
13. *What are the risk factors for stroke in patients with monocular visual symptoms?*
    - Risk factors include:[8]
      - Age > 70 years
      - Male gender
      - History of TIA
      - History of intermittent claudication
      - Ipsilateral stenosis (80–99%)
      - Number of collaterals on angiography
14. *How would you treat this patient?*
    - The patient was treated with excision of the stump and common carotid artery (CCA) endarterectomy and patch angioplasty with only minor improvement in symptoms.
    - ▶**Fig. 51.3** presents a treatment algorithm for patients with ophthalmic manifestations of carotid disease.

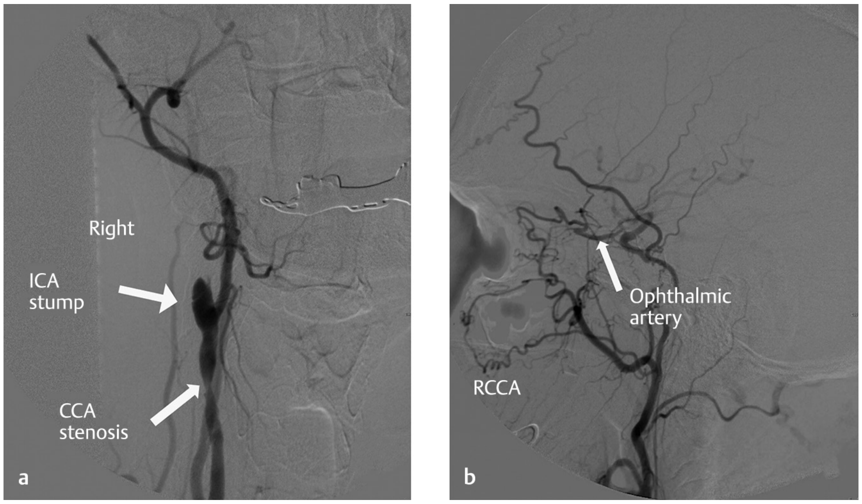

**Fig. 51.2** Angiogram: (**a**) Right common carotid artery (RCCA) injection showing internal carotid artery (ICA), distal stump (*arrow*), and RCCA stenosis (*arrow*)**.** (**b**) Selective external carotid artery injection showing communication between the internal and external circulation via the ophthalmic artery (*arrow*)**.**

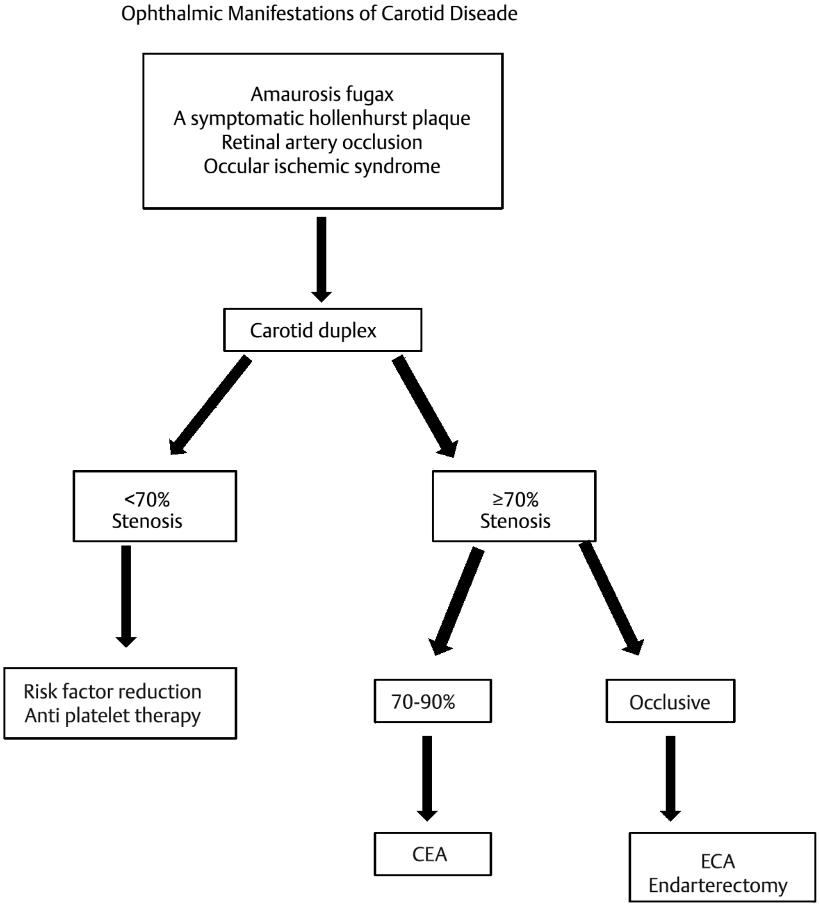
Ophthalmic Manifestations of Carotid Diseade
Amaurosis fugax
A symptomatic hollenhurst plaque
Retinal artery occlusion
Occular ischemic syndrome
Carotid duplex
<70%
Stenosis
≥70%
Stenosis
Risk factor reduction
Anti platelet therapy
70-90%
Occlusive
CEA
ECA
Endarterectomy

**Fig. 51.3** Treatment algorithm for patients with ophthalmic manifestations of carotid disease. (Data from Wolintz 2005).[11]

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# Case 52 Tandem Extracranial and Intracranial Carotid Stenosis

*Glenn C. Hunter and Craig C. Weinkauf*

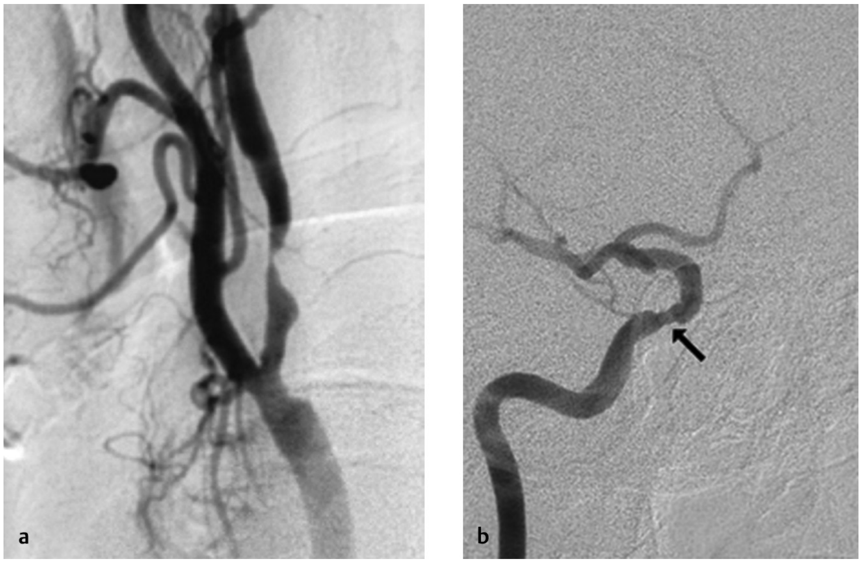

**Fig. 52.1** Selective cerebral angiogram demonstrating (**a**) 70% extracranial ICA stenosis, and (**b**) 50% intracranial ICA stenosis.

## Clinical Presentation

- A 67-year-old right-handed female presents to the emergency room (ER) with recurrent episodes of left hemispheric transient ischemic attacks (TIAs) that present with speech difficulty and right-sided weakness.
- She has a history of hypertension, hyperlipidemia, type 2 diabetes mellitus, and had undergone a femoral-popliteal bypass 2 years prior to this presentation.
- Pulse rate is 106 beats per minute, irregular. Blood pressure is 140/95 mm Hg and there are bilateral carotid bruits.
- Neurologic exam was intact.

## Questions

1. What is the differential diagnosis?
2. The ER physician obtains a CT scan, electrocardiogram (EKG), and orders a carotid duplex exam. Are there any further tests you would obtain at this time?

   The EKG shows atrial fibrillation and duplex scan shows 50–79% bilateral internal carotid artery (ICA) stenosis.
3. What are the criteria for hospital admission of this patient?
4. What is your initial management of this patient?

   The patient has urgent family problems and does not want to be admitted to hospital. She is already on aspirin so you add clopidogrel. Two days later, she returns to the ER with another TIA that lasted 2 hours.
5. How does a recurrent TIA change your management and what imaging study would you order?

   A four-vessel cervical and cerebral angiogram is ordered. The angiogram shows 70% bilateral ICA stenosis with a 70% siphon stenosis of the ICA (**Fig. 52.1**). The left A-1 segment is not well visualized.
6. What are the risk factors for intracranial atherosclerotic disease (ICAD)? What are the risk factors for concurrence of extracranial atherosclerotic disease (ECAD) and ICAD disease?
7. List the causes of symptoms in patients with tandem carotid stenosis.
8. What is the usual distribution of intracranial lesions?
9. What is the added risk of the presence of ICAD in cases of preexisting ECAD?

## Questions (*continued*)

10. Discuss the recommended treatment for combined disease.
11. What is the risk of stroke, in general, in patients with carotid stenosis both symptomatic and asymptomatic? Provide your answer based on percentage of stenosis and modality of treatment (surgical vs. medical).
12. When would you perform surgery on this patient if she had had a stroke rather than a TIA?
13. What is the management of patients when they are not surgical candidates?

## Answers

1. *What is the differential diagnosis?*
   The differential diagnosis should include the following:
   - Carotid stenosis
   - Cardiac embolism
   - Carotid dissection
   - Hemiplegic migraine
   - Seizure disorder
   - Subdural hematoma
   - Tumor
   - Hypo/hyperglycemia
2. *The ER physician obtains a CT scan, electrocardiogram (EKG), and orders a carotid duplex exam. Are there any further tests you would obtain at this time?*
   - Complete blood count
   - Screen electrolytes/renal function tests
   - Blood glucose, serum lipids
   - Cardiac enzymes
   - Coagulation studies
   - C-reactive protein/erythrocyte sedimentation rate (ESR)
   - Autoimmune vasculitis screen
   - Chest X-ray

   The EKG shows atrial fibrillation and duplex scan shows 50–79% bilateral ICA stenosis.
3. *What are the criteria for hospital admission of this patient?*
   - Hospital admission should be considered for patients who present within 24–48 hours of their first TIA with:
     - Crescendo TIAs
     - Symptoms of > 1 hour's duration
4. *What is your initial management of this patient?*
   - Initial management of the patient should include:
     - Admission to hospital for control of blood pressure, glucose, and arrhythmias
     - Consider intravenous (IV) heparin if there is no intracranial bleed
     - MRI/MR angiography (MRA) or CT angiography (CTA)[1]
     - Cardiac echocardiography
5. *How does a recurrent TIA change your management and what imaging study would you order?*
   Following steps should be taken in case of a recurrent TIA:
   - Direct hospital admission
   - Repeat CT/MRI
   - Repeat duplex scan
   - Transcranial Doppler[2]
   - A CTA, MRA, or digital subtraction angiogram (DSA) is considered. CTA is the usual screening test prior to doing an angiogram.

   A four-vessel cervical and cerebral angiogram is ordered. The angiogram shows bilateral 70% ICA stenosis with a 50% carotid siphon stenosis of the left ICA (**Fig. 52.1**). The left A-1 segment is poorly visualized.
6. *What are the risk factors for intracranial atherosclerotic disease (ICAD)? What are the risk factors for concurrence of extracranial atherosclerotic disease (ECAD) and ICAD disease?*
   Risk factors for ICAD are as follows:[3–5]
   - Race: African American or Asian[3]
   - Gender: risk of 28.4% in females versus 16.6% in males[4]
   - Hypertension
   - Hyperlipidemia
   - Diabetes mellitus

   Risk factors for combined ECAD and ICAD include:[2,6]
   - Diabetes mellitus
   - Coronary artery disease
7. *List the causes of symptoms in patients with tandem carotid stenosis.*
   Etiology of symptoms in patients with tandem disease includes:
   - Embolism from carotid bifurcation/less commonly from the ICAD lesion
   - Ipsilateral carotid occlusion
   - Hypoperfusion-ICAD > ECAD
8. *What is the usual distribution of intracranial lesions?*
   Distribution of ICAD
   - ICA: 49%
   - Middle cerebral artery (MCA): 20%
   - Posterior cerebral artery (PCA): 11%
   - Vertebrobasilar artery (VB): 1.1%
   - Anterior communicating artery (ACOM): 9%
9. *What is the added risk of the presence of ICAD in cases of preexisting ECAD?*
   Presence of ICAD in cases of preexisting ECAD adds following risks:
   - Deficits persist longer
   - Plaques more fibrotic[7]
   - Independent risk factor for stroke[3,8]

## Answers (*continued*)

- Risk of stroke is greater in ICAD as compared with MCA lesions alone (36 vs. 24%)[2,3,5,6,8]

10. *Discuss the recommended treatment for combined disease.*

    Treatment of combined ECAD and ICAD stenosis
    - Carotid endarterectomy (CEA) in cases of 70–90% stenosis[3]
    - CEA if 50–79% ECAD stenosis and 70% ICAD stenosis[2,3,5]
    - Other options include:
      - CEA and ICAD angioplasty; (Angioplasty and stenting are increasingly being used for intracranial disease. Long-term results and large population studies are not yet available.)[6]
      - Internal carotid artery stenting (CAS)[6] and ICAD angioplasty (32% restenosis at 6 months)
      - Clot thrombectomy and stenting may benefit acute extracranial carotid artery occlusion with ICAD.[9–11]
      - CEA is not beneficial if the ICAD is worse than the ECAD.
11. *What is the risk of stroke, in general, in patients with carotid stenosis both symptomatic and asymptomatic? Provide your answer based on percentage of stenosis and modality of treatment (surgical vs. medical).*
    - See ▶**Table 52.1**[2,3,5,6] and ▶**Table 52.2**[6,9–14] for a summary.
    - In symptomatic cases:
      - CEA is of benefit if there is 70–99% stenosis as long as surgical morbidity and mortality (M&M) is ≤ 6%. The stroke risk is overall decreased by ~8% per year.[9,10]
      - CEA is of marginal benefit in cases of 50–69% stenosis. Consider this procedure if there are associated risk factors such as:
        - An associated ulcerative lesion[7] (as in this case)
        - Contralateral ICA occlusion
        - Male gender
        - Intraluminal thrombus
        - Stroke more than transient (not TIA)
        - Younger age at presentation
      - In asymptomatic cases, CEA is of marginal benefit in cases of 60–99% carotid stenosis cases.[11,12]
      - Consider performing this procedure if:
        - The overall operative risk (M&M) is ~3% or less.
        - There are associated risk factors such as the ones described above.
12. *When would you perform surgery on this patient if she had had a stroke rather than a TIA?*

    The timing of an intervention depends on the acuity of the stroke and the extent of the neurologic deficit. If the patient is seen within 6 hours, consideration should be given to emergent thrombolysis/thrombectomy and stenting.[8,14,–21] Surgery is otherwise delayed until the symptoms resolve and the deficits have plateaued. Use of a shunt during CEA should be considered in patients who have had a stroke.
13. *What is the management of patients when they are not surgical candidates?*

    The observed 5-year survival rate of patients with ICAD is 60% compared to an expected rate of 87% in the normal population (50% of deaths are cardiac related).[21] The management other than surgery includes:
    - Medical therapy for patients with inoperable disease
    - Control: blood pressure, blood sugar, lipids
    - Warfarin (Coumadin; Bristol-Myers Squibb, New York, NY)

**Table 52.1** Comparison or risks of stroke between medical and surgical treatment modalities in patients with symptomatic carotid stenosis (estimates based on NASCET and ECST studies)

| Stenosis | Risk of stroke at 2 years with medical therapy (%) | Risk of stroke at 2 years with surgical therapy (CEA) (%) | Absolute risk reduction (%) | Perioperative M&M (%) |
|---|---|---|---|---|
| 70–99% NASCET (8) | 25 | 9 | 16 | 6 |
| 50–69% NASCET (8) | 14.5 | 9.5 | 5 | 6 |
| 50–69% ESCT (10) | 9.7 | 11 | −1.5 | 10 |

*Abbreviations:* CEA, carotid endarterectomy; ECST, European Carotid Surgery Trial; M&M, morbidity and mortality; NASCET, North American Symptomatic Carotid Endarterectomy Trial.
*Source:* From North American Symptomatic Carotid Endarterectomy Trial Collaborators 1991; Randomised trial of endarterectomy for recently symptomatic carotid stenosis: final results of the MRC European Carotid Surgery Trial (ECST) 1998; Mohr et al 2004.[9,10,17]

**Table 52.2** Comparison or risks of stroke between medical and surgical treatment modalities in patients with asymptomatic carotid stenosis (estimates based on ACAS and VA studies)

| Stenosis | Risk of stroke with medical therapy (%) | Risk of stroke with surgical therapy (%) | Absolute risk reduction (%) | Perioperative M&M (%) |
|---|---|---|---|---|
| 60–99% ACAS[6] | 11 | 5 | 6 | 1.2 |
| RISK at 5 years | | | | |
| 50–99% VA study[9] | 10 | 5 | 5 | 4 |
| RISK at 4 years | | | | |

*Abbreviations:* ACAS, Asymptomatic Carotid Atherosclerosis Study; CEA, carotid endarterectomy; M&M, morbidity and mortality; VA, Veterans Affairs.
*Source:* From Executive Committee for the Asymptomatic Carotid Atherosclerosis Study 1995; Hobson et al 1993; Findlay et al 2004.[11,12,18]

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# Case 53 Vertebral Artery Stenosis with Ischemia

*Glenn C. Hunter and Rudiger von Ritschl*

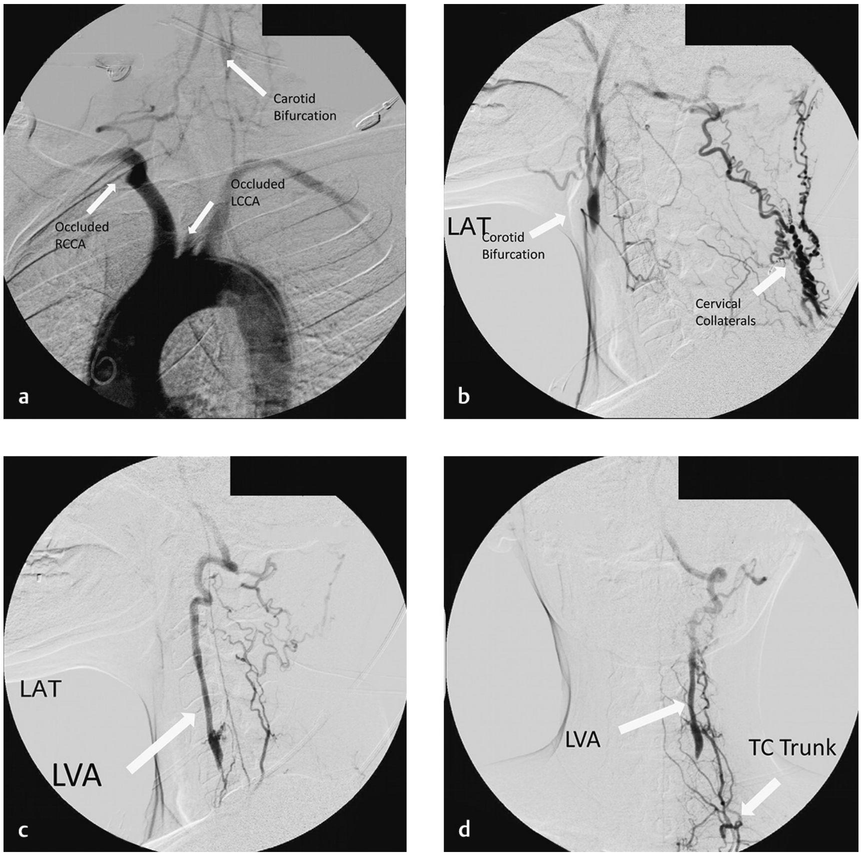

**Fig. 53.1** Arch aortogram showing the origins of the innominate, left carotid, left subclavian, and vertebral arteries (**a**). Delayed distal views demonstrating the left carotid bifurcation (**b**) and left vertebral artery (LVA) reconstituted via cervical collaterals (**c**) and thyrocervical (TC) trunk (**d**). LAT, lateral thoracic artery; LCCA, left common carotid artery; RCCA, right common carotid artery.

## Clinical Presentation

- A 56-year-old man with hypertension and type 2 diabetes mellitus presents with a 6- to 9-month history of inability to raise his head or get out of bed without symptoms of lightheadedness, dizziness, blurred vision, and drop attacks. His symptoms have become so severe that he is currently bedbound. He is taking aspirin and antihypertensive agents.
- Physical examination revealed a healthy appearing man. His blood pressure is 150/90 mm Hg on the right and 135/80 mm Hg on the left; there are bilateral carotid bruits.

## Questions

1. What is the differential diagnosis?
2. What tests would you obtain?
3. Based on the findings of the duplex scan (bilateral internal carotid artery [ICA] occlusion with no antegrade flow in either VA), how would you proceed? Is the absence of antegrade flow always indicative of VA occlusion?
4. You have a choice of ordering CT angiography (CTA), MR angiography (MRA), or a digital subtraction cerebral angiogram (DSA) to evaluate the extracranial (EC) and intracranial (IC) vasculature. What factors should you consider in making your decision?

   With the extent of this patient's EC occlusive disease, one would assume that this patient's symptoms were most likely due to global cerebral hypoperfusion.
5. Is this assumption correct in all patients with vertebrobasilar insufficiency (VBI)? What are the most frequent causes of symptoms in patients with VBI?

## Questions (*continued*)

6. A four-vessel angiogram is obtained. Describe the angiographic findings (▶**Fig. 53.1**).
7. How would your management differ if the angiogram showed unilateral or bilateral 70–99% ICA or CCA stenosis rather than occlusion of both ICAs or CCAs, in addition to bilateral VA occlusion? How does the presence or absence of symptoms influence your decision?
8. What operative procedures are performed for VBI?
9. What alternative interventional therapies are available?
10. What are the main comorbidities and the medical management of patient with VBI?
11. How would you treat this patient?
12. VA stenting is gaining increasing popularity to treat patients with VBI. How do the outcomes of patients undergoing VA stenting fair in comparison with those receiving best medical therapy?

## Answers

1. *What is the differential diagnosis?*
   - VBI
   - Postural hypotension
   - Vertigo
   - Cardiac arrhythmias/aortic arch thromboembolism
   - Autoimmune vasculitis
   - Hypoglycemia
   - Hypovolemia
   - Hyperventilation
2. *What tests would you obtain?*
   - Cardiac evaluation: electrocardiogram (EKG), Holter monitoring, and transesophageal echocardiography (suspected cardiac or aortic arch disease)
   - Blood chemistries: electrolytes, blood glucose, complete blood count (CBC), coagulation profile, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and autoimmune work-up
   - Vascular imaging studies: carotid duplex scan and transcranial Doppler
   - Cerebral imaging studies: MRI, MRA, CTA, or DSA
   - ENT exam (when indicated)
3. *Based on the findings of the duplex scan (bilateral internal carotid artery [ICA] occlusion with no antegrade flow in either VA), how would you proceed? Is the absence of antegrade flow always indicative of VA occlusion?*
   The absence of antegrade flow on a duplex scan is not diagnostic of VA occlusion. Anatomic variations including an arch origin and hypoplasia of the VAs may account for the absence of antegrade VA flow.
4. *You have a choice of ordering CT angiography (CTA), MR angiography (MRA), or a digital subtraction cerebral angiogram (DSA) to evaluate the extracranial (EC) and intracranial (IC) vasculature. What factors should you consider in making your decision?*
   - CTA can be used to assess the carotid arteries (CAs) and VAs in the neck. The use of ionizing radiation, the potential for contrast-induced nephrotoxicity, interference from boney artifacts in the cervical spine, and vessel wall calcification, especially at the origins of the VAs limits the use of CTA. The next generation of multisector CT scanners, may potentially improve spatial resolution.
   - Contrast-enhanced MRA (CEMRA) allows assessment of both EC and IC anterior and posterior circulation occlusive disease and is superior to time-of-flight MRA. Studies comparing CEMRA and DSA have shown equivalent sensitivity and specificity rates.
   - DSA: In this patient, arch aortography with selective catheterization of the innominate and left subclavian arteries with delayed image acquisition was used to evaluate the extent of the occlusive disease of the EC and IC carotid and vertebral arteries. CEMRA would have been an alternative choice if it was available when this patient was treated.

   *With the extent of this patient's EC occlusive disease, one would assume that this patient's symptoms were most likely due to global cerebral hypoperfusion.*
5. *Is this assumption correct in all patients with vertebrobasilar insufficiency (VBI)? What are the most frequent causes of symptoms in patients with VBI?*
   This patient had occlusion of all four vessels supplying blood to the brain; therefore, it is quite likely that his symptoms were due to global hypoperfusion. However, most patients with VBI do not have diminished cerebral blood flow.
   - The major causes of VBI are listed below in order of frequency:
     - Large vessel EC and IC occlusive disease: 41%
     - Hemodynamic causes: 33%
     - Cardiac or aortic arch embolism: 24%
6. *A four-vessel angiogram is obtained. Describe the angiographic findings (*▶**Fig. 53.1**).
   Arch aortogram shows the origins of the innominate, left carotid, left subclavian arteries and VAs. There is occlusion of the right and left common carotid arteries (CCAs) and both VAs at their origins. Delayed acquisition images demonstrate reconstitution the left carotid bifurcation (B) and left VA (LVA) via cervical collaterals (C) and (D) thyrocervical trunk.

## Answers (*continued*)

7. *How would your management differ if the angiogram showed unilateral or bilateral 70–99% ICA or CCA stenosis rather than occlusion of both ICAs or CCAs, in addition to bilateral VA occlusion? How does the presence or absence of symptoms influence your decision?*
   The efficacy of treating VA occlusion with concomitant symptomatic or asymptomatic carotid occlusive disease remains controversial. However, if there is unilateral or bilateral symptomatic or asymptomatic high-grade (70–99%) ICA stenosis, then carotid endarterectomy (CEA) or carotid stenting (CS) should be performed prior to treatment of the VA occlusion. The symptomatic high-grade ICA stenotic lesion should be treated first: if the patient is asymptomatic, blood flow is restored to the dominant hemisphere first. CCA stenosis/occlusion is much less common than the ICA stenosis (10–15% vs. 85–90%) but similar principles apply.
8. *What operative procedures are performed for VBI?*
   - CEA to provide antegrade flow to the anterior circulation.
   - Carotid subclavian bypass: the inflow may originate from the subclavian artery if the CCA is occluded and the carotid bifurcation is patent as in this patient. If the proximal subclavian is occluded, the bypass will provide antegrade flow from the CCA.
   - Subclavian or VA endarterectomy; and subclavian–VA bypass saphenous vein bypass: these procedures are technically difficult and should only be undertaken by surgeons experienced in these techniques.
9. *What alternative interventional therapies are available?*
   - EC carotid angioplasty and stenting
   - Subclavian artery angioplasty and stenting
   - EC, IC, and VA angioplasty and stenting
   - Thrombectomy or thrombolysis (controversial in posterior circulation)
10. *What are the main comorbidities and the medical management of patient with VBI?*
    - Comorbidities include: hypertension (61%), diabetes mellitus (26%) smoking (36%), hyperlipidemia (25%), and coronary (35%) and peripheral vascular occlusive disease (10%)
    - Medical management includes:
      - Optimization of blood pressure control
      - Controlling the risk factors for atherosclerosis
      - Warfarin (Coumadin; Bristol-Myers Squibb, New York, NY) for obstructive lesions not amenable to surgery or endovascular therapy
      - Thrombectomy/thrombolysis
      - Aspirin/clopidogrel (Plavix; Bristol-Myers Squibb, New York, NY) after stenting
      - Tissue plasminogen activator (tPA)/abciximab
11. *How would you treat this patient?*
    This patient was treated with a left subclavian to carotid bifurcation bypass using a polytetrafluoroethylene (PTFE) graft and a standard endarterectomy with patch angioplasty of the bifurcation and ICA with resolution of his symptoms.
12. *VA stenting is gaining increasing popularity to treat patients with VBI. How do the outcomes of patients undergoing VA stenting fair in comparison with those receiving best medical therapy?*
    - In the Stenting of Symptomatic Atherosclerotic Lesions in the Vertebral or Intracranial Arteries (SSYLVIA) Study, 18 (29.5%) EC VAs (6 ostia, 12 proximal to the posterior inferior cerebellar artery [PICA]) were treated. Strokes occurred in 6.6% of patients within 30 days and in 7.3% patients between 30 days and 1 year. Restenoses occurred in 35% of patients, 61% were asymptomatic. Because of the heterogeneous mix of patients in this study it is difficult to discern the complication rate for each vascular territory treated.
    - Results of the Veterans Administration Acute Stroke (VAST) study show that the risk of recurrent VB stroke is low in patients with best medical therapy whereas VA stenting is associated with a major periprocedural vascular complication in about 1:20 patients. The conclusions of this randomized study show that VA stenting offers no significant therapeutic benefit over best medical therapy.

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# Case 54 High-Grade Carotid Stenosis and Intracranial Aneurysm

*Glenn C. Hunter and Remi Nader*

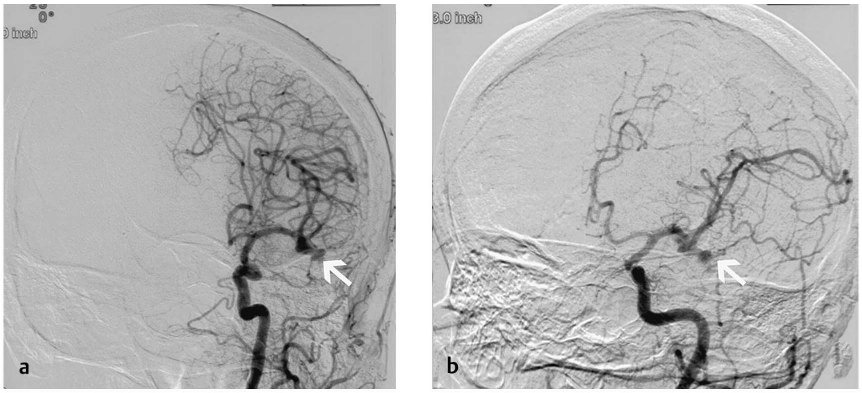

**Fig. 54.1** Cerebral angiogram, left carotid injection, showing a 7-mm left middle carotid artery trifurcation aneurysm on (**a**) anteroposterior and (**b**) lateral views.

## Clinical Presentation

- A 74-year-old woman presents with a history of left hemispheric transient ischemic attacks (TIAs).
- She experienced one episode of right arm and leg weakness about 4 weeks ago that completely resolved within 5 to 7 minutes.
- Her blood pressure is 160/110 mm Hg on two antihypertensive medications. The blood pressure in her left arm is 30 mm Hg lower than that on the right side.
- Bilateral carotid bruits are present.

## Questions

1. What initial studies would you order?
2. What additional imaging studies would assist in the diagnosis?
3. How would you manage this patient initially?

   A carotid duplex scan shows a high-grade (80–99%) left internal carotid artery (LICA) stenosis with 50 to 79% stenosis on the right side.
   The patient is sent home on aspirin and clopidogrel (Plavix; Bristol-Myers Squibb, New York, NY), but returns 2 weeks later with complaints of having discontinued her medication because of bruising. She has experienced two episodes of TIAs since discontinuing clopidogrel.

4. How do you manage her now?
5. A digital subtraction cerebral angiogram (DSA) is obtained. Is this study preferable to CT angiography (CTA) or MR angiography (MRA)?

   The angiogram demonstrated an irregular high-grade (90%) LICA stenosis (not shown here) and a left 7-mm middle cerebral artery aneurysm (▶**Fig. 54.1**).

6. What is the prevalence and sex distribution of an unruptured intracranial aneurysm (UIA)?
7. What is the incidence of an UIA on cerebral angiograms performed for carotid disease?
8. Are there any conditions associated with intracranial aneurysms?
9. How may intracranial aneurysms present?
10. What are the factors that predispose to aneurysm rupture?
11. What is the estimated risk of rupture in this case?
12. List the therapeutic options for this patient.
13. Outline the factors that need to be considered in selecting the treatment option.
14. Describe the anatomical segments of the ICA and its branches.

## Answers

1. *What initial studies would you order?*
   - The patient should have an initial work-up for stroke.
   - Laboratory studies should include: complete blood count (CBC), coagulation studies, blood sugar, electrolytes, renal function tests, erythrocyte sedimentation rate, C-reactive protein
   - Chest X-ray and electrocardiogram (EKG)
2. *What additional imaging studies would assist in the diagnosis?*
   - A carotid duplex scan
   - CT or MRI of the brain

   A repeat duplex scan (to exclude interval carotid occlusion) confirms 80 to 99% LICA stenosis with 50 to 79% stenosis on the right side.

   A nonenhanced CT scan shows no evidence of hemorrhage or ischemia.
3. *How would you manage this patient initially?*
   - She was placed on aspirin and clopidogrel.
   - Arrangements should be made for an outpatient vascular imaging study (CTA, MRA, or DSA).
   - Because this was her first TIA, it is important to ensure she is on adequate antiplatelet therapy and it is acceptable to order her imaging studies as an outpatient.
4. *How do you manage her now?*
   - She is experiencing more frequent TIAs; therefore, direct hospital admission is indicated.
   - Consider intravenous heparin therapy as no hemorrhage was seen on the CT scan of the brain.
5. *A digital subtraction cerebral angiogram (DSA) is obtained. Is this study preferable to CT angiography (CTA) or MR angiography (MRA)?*
   - Because of the rapid acquisition of images and availability, a CTA is almost invariably obtained as a screening exam in both symptomatic and asymptomatic patients with these duplex findings. However, calcification, ossification, and some types of hardware may obscure visualization.
   - MRA allows more options for image contrast and is less sensitive to calcification, ossification, and some types of hardware.
   - Both CTA and MRA are good for diagnosing normal arteries and severely stenotic lesions. MRA has good negative predictive value for excluding 50 to 99% but has a low positive predictive value.
   - This patient has evidence of both left subclavian and bilateral carotid stenosis. A DSA was obtained. The DSA allows good spatial and temporal resolution of individual vessels and is both diagnostic and interventional. The minor risk of local complications, the dangers of ionizing radiation and contrast media should be discussed with the patient prior to the study.
6. *What is the prevalence and sex distribution of an unruptured intracranial aneurysm (UIA)?*
   - The prevalence of UIA ranges from 0.8 to 8% (mean 5%).
   - There is a 2:1 female-to-male ratio.[1]
   - Incidence increases with age.[1]
7. *What is the incidence of an UIA on cerebral angiograms performed for carotid disease?*
   - Incidence: 1/40 patients with symptomatic carotid stenosis have UIAs.[2] Based on the North American Symptomatic Carotid Endarterectomy Trial (NASCET) study,[3] there is a 3.1% incidence of an UIA in cerebral angiograms performed for carotid disease.
     - Ninety-six percent of aneurysms are less than or equal to 10 mm in size.
     - Fifty-five percent are ipsilateral to the side of the carotid stenosis.
     - These account for 6 to 16% of subarachnoid hemorrhages (SAHs)
     - Eight to 34% UIAs are multiple.
8. *Are there any conditions associated with intracranial aneurysms?*
   - UIAs can have a congenital predisposition and can be associated with following:[1]
     - Aortic coarctation
     - Polycystic kidneys (autosomal dominant)
     - Family history of UIA/SAH
     - Fibromuscular dysplasia
     - Pseudoxanthoma elasticum
     - Moyamoya disease
     - Systemic lupus erythematosus
     - Arteriovenous malformations
9. *How may intracranial aneurysms present?*
   - Incidental finding
   - SAH
   - Embolism (rare)
   - Headache
   - Cranial nerve palsy[1]
10. *What are the factors that predispose to aneurysm rupture?*
    - Size
    - Location (anterior vs. posterior circulation)
    - Bleb formation
    - Previous SAH (increases hemorrhage rate by 11-fold)[4]
11. *What is the estimated risk of rupture in this case?*
    - Risk of rupture in this case is less than 1%. Rupture risk depends on size and location of the aneurysm.[1,4–6]
    - See prior aneurysm cases for a detailed description of the associated risks.
12. *List the therapeutic options for this patient.*
    - Carotid endarterectomy (CEA) without UIA clipping (UIA observation)
    - Combined CEA and UIA clipping
    - CEA and aneurysm coiling
    - Carotid artery stenting (CAS) and aneurysm coiling[7]

## Answers (*continued*)

13. *Outline the factors that need to be considered in selecting the treatment option?*
    - Determine which lesion is causing the symptoms.
    - Determine the relationship between the two lesions:
      - Embolic symptoms are most often due to carotid bifurcation plaque.
      - Carotid lesions are a more common cause of symptoms (as in this case).
    - Some factors with the treatment of one lesion may exacerbate the risks of the other:
      - CEA may increase the risk of aneurysm rupture due to an increase in cerebral perfusion pressure (this is very unlikely, however).[8]
      - Hypotension necessary to clip an aneurysm may cause thrombosis of a high-grade carotid stenosis.[8]

    The occurrence of high-grade EC stenosis with UIAs gives rise to several therapeutic dilemmas. There is some evidence to suggest that there is a higher prevalence of UIAs with carotid occlusive disease. There is also the concern that EC revascularization may increase the risk of aneurysm growth and rupture. Current evidence from the literature suggests that the risk of aneurysm growth and rupture following an EC intervention is low, and therefore, EC revascularization can be recommended in the majority of patients with a favorable risk profile having high-grade EC stenosis and UIAs of ≤ 10 mm. However, there are no prospective studies addressing this problem.[9–12]
    - Based on the NASCET study,[3] the 5-year risk of rupture of UIA differs as per the treatment modality of the underlying carotid disease.
      - With CEA, it is 10%
      - With best medical therapy, it is 22.7%
14. *Describe the anatomical segments of the ICA and its branches.*
    - See ▶**Fig. 54.2** for a detailed description.

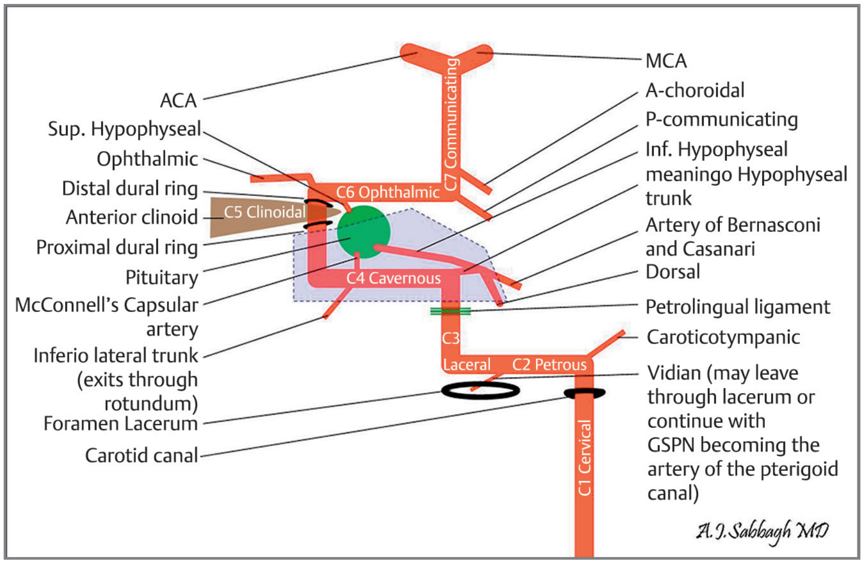
MCA
A-choroidal
P-communicating
Inf. Hypophyseal
meaningo Hypophyseal trunk
Artery of Bernasconi and Casanari
Dorsal
Petrolingual ligament
Caroticotympanic
Vidian (may leave through lacerum or continue with GSPN becoming the artery of the pterigoid canal)
ACA
Sup. Hypophyseal
Ophthalmic
Distal dural ring
Anterior clinoid
Proximal dural ring
Pituitary
McConnell's Capsular artery
Inferio lateral trunk (exits through rotundum)
Foramen Lacerum
Carotid canal
C7 Communicating
C6 Ophthalmic
C5 Clinoidal
C4 Cavernous
C3 Laceral
C2 Petrous
C1 Cervical
A.J. Sabbagh MD

**Fig. 54.2** Anatomical segments of the internal carotid artery and their branches. A, anterior; ACA, anterior cerebral artery; GSPN, greater superficial petrosal nerve; Inf, inferior; MCA, middle cerebral artery; P, posterior; Sup, superior.

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# Section III Intracranial Pathology: Cranial Trauma

# Case 55 Chronic Subdural Hematoma

*Remi Nader*

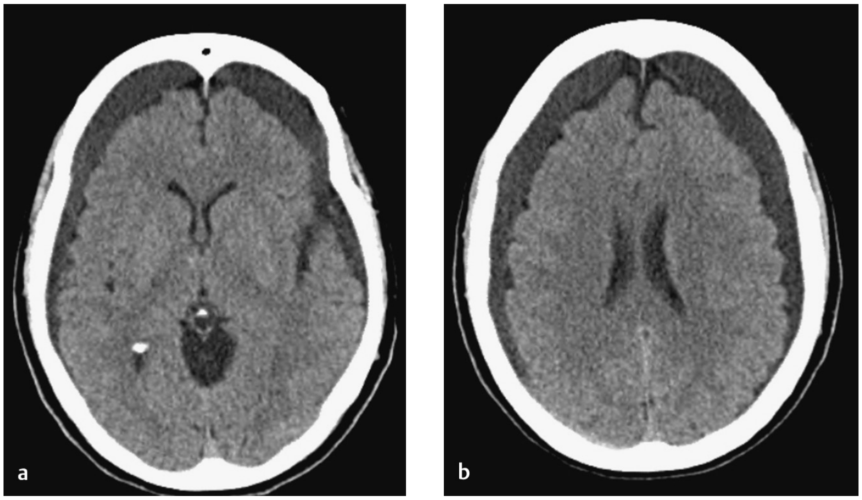

**Fig. 55.1** (**a, b**) CT scan of the brain without contrast.

## Clinical Presentation

- A 70-year-old woman with end-stage Alzheimer's disease, diabetes mellitus, nursing home bound, is brought to the emergency room (ER) for a 3-day episode of increasing confusion, agitation, and inappropriate behavior.
- The patient is confused at baseline; however, she was noted to be combative and refusing her care as well as voicing aloud her request to leave the nursing home and to go home, which is unusual behavior for her.
- She is a poor historian, but she does report having had frequent falls in the near past.
- Physical examination reveals a left-sided pronator drift and confusion. The remainder of the examination is normal.
- CT scan is obtained and shown in ▶**Fig. 55.1**.

## Questions

1. Interpret the CT scan.
2. What other questions or information would you like to obtain to further plan your management?
3. What further studies or investigations would you like to obtain?
4. What would you tell the family, next of kin, or caregiver?

   The patient is not on any anticoagulant or anti-inflammatory medications. Her laboratory results, including complete blood count, electrolytes, and coagulation profile, are within normal limits. You had a discussion with the family about the treatment options and they opted for surgery.

5. How would you approach this case surgically? Describe all the steps of surgery including positioning, incision, bone opening, evacuation, and closure.
6. Preoperatively, as you plan the surgery, she develops hyponatremia with sodium level of 118 mEq/L, and she becomes more lethargic. What are some causes of hyponatremia and which is the most likely in this case?
7. How would you manage this problem?

   She does well from bilateral burr hole evacuation of subdural hematomas and is discharged home. However, she comes back 3 weeks later to your clinic. She now has some purulent discharge from the right posterior incision. She is somewhat somnolent and has a left hemiparesis slightly worse than the one she had immediately postoperatively.

8. What is your course of action now?
9. A follow-up CT scan and subsequently an MRI scan are obtained (▶**Fig. 55.2**). Interpret the studies and provide a differential diagnosis.
10. What is your management plan?

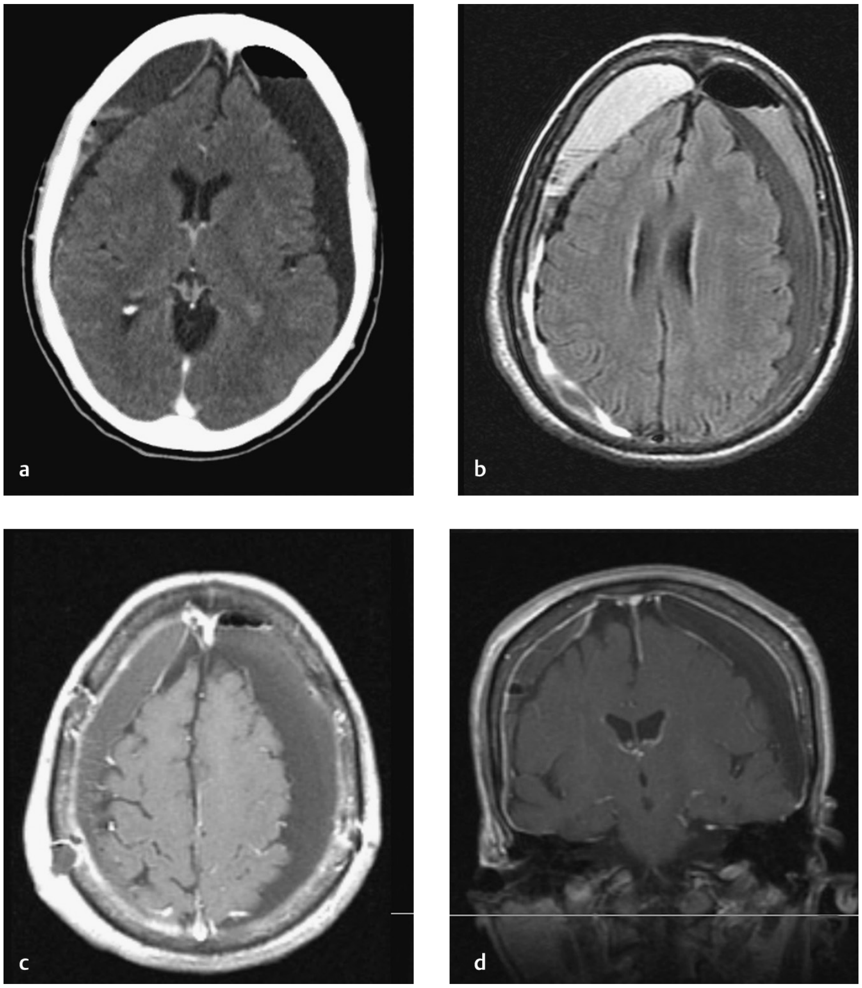

**Fig. 55.2** (**a**) CT scan with contrast and (**b**) MRI fluid-attenuated inversion recovery axial image without contrast, and T1-weighted images with contrast, (**c**) axial and (**d**) coronal.

## Answers

1. *Interpret the CT scan.*
   - CT scan of the brain reveals bilateral subdural hematomas which are mainly chronic with a very small subacute component on the right side.
   - The right subdural hematoma is ~1.5 cm in thickness. The left subdural hematoma is ~1.3 cm in thickness. They are both mainly along the convexity in the frontal and somewhat parietal area.
   - There is some diffuse brain atrophy in both frontal lobes consistent with advanced Alzheimer's disease.
   - There also appears to be no midline shift, no obvious mass effect, and no edema. The basal cisterns are wide open.
   - There might be some effacement in the sulci along the cortical surface in the frontal lobes.
2. *What other questions or information would you like to obtain to further plan your management?*
   - Obtain current medication history, especially whether or not she is on anticoagulants.
   - Determine if there is a living will or a durable power of attorney. If so, determine the patient's wishes in a situation where lifesaving measures are needed.
3. *What further studies or investigations would you like to obtain?*
   - Laboratory tests: complete blood count, electrolytes, coagulation profile, type and screen
   - Other imaging modalities: MRI—unlikely to change your management in this case
   - If surgical intervention is contemplated, then medical clearance may be needed (electrocardiogram [EKG], chest X-ray, cardiac echography, etc.)
4. *What would you tell the family, next of kin, or caregiver?*
   - The patient has fluid collections on the surface of her brain which are likely due to old hemorrhages.
   - It is not clear whether or not these fluid collections are causing a lot of pressure on her brain.
   - She also has brain atrophy due to Alzheimer's disease.
   - She may possibly benefit from evacuation of the fluid collections.[1]
   - However, the caregiver needs to be aware of the fact that evacuation may also not help as she may be

## Answers (*continued*)

having progressive dementia due to Alzheimer's and she may also have complications from the surgery.

- Inform the caregiver of all the potential surgical complications which include, but are not limited to, pain; bleeding; infection; failure to treat presenting condition; need for further surgeries or procedures in the future; reaccumulation of the fluid collections causing damage to adjacent blood vessels, nerves, and tissues; additional loss of brain function such as memory, stroke, changes in vision, deafness, inability to smell, coordination loss, seizures, cerebrospinal fluid leak; weakness; impaired muscle function; numbness; paralysis; and death.[2–4]

5. *How would you approach this case surgically? Describe all the steps of surgery including positioning, incision, bone opening, evacuation, and closure.*
   - One operative option consists of bilateral burr hole evacuation of the subdural hematoma, with two burr holes placed on each side.[5–9]
   - Preoperative preparation includes:
     - Preoperative antibiotics: cefazolin 1 g intravenously (IV) every 8 hours
     - Preoperative seizure prophylaxis: phenytoin, loading dose of 1 g IV over 1 hour, followed by a maintenance dose of 100 mg every 8 hours
     - Positioning: supine with head of bed slightly elevated
     - Anesthesia: preferably conscious sedation or general endotracheal if the patient will tolerate it (i.e., no cardiac or pulmonary risk factors)
   - Surgical steps include:
     - Clip the hair and mark incisions along the convexity in-line with each other such that if the need for a bone flap arises it will suffice to connect both incisions.
     - The incisions should be ~5 to 6 cm away from the midline; one along the parietal boss and one just behind the hair line anteriorly—both ~3 cm in length.
     - Start by opening the right side as this is the symptomatic side.
     - Open skin, galea, and periosteum with a no. 10 blade; expose the skull and make a burr hole ~2.5 cm in diameter with a pneumatic drill.
     - Wax the bone edges and cauterize the dura.
     - Open the dura in cruciate fashion, then drain the blood.
     - Irrigate both burr holes profusely with normal saline until the drainage becomes clear.
     - One may place a red rubber catheter in the burr hole subdurally and further irrigate to dislodge any pockets of blood that are harder to access.
     - Closure is completed by placing a piece of Gelfoam (Pfizer, Inc., New York, NY) on top of the burr hole and closing the skin and galea with 3–0 Vicryl (Ethicon, Somerville, NJ) and staples.
     - One may leave a small drain in the subgaleal space.
   - The patient is kept with the head of the bed flat postoperatively.[10]
6. *Preoperatively, as you plan the surgery, she develops hyponatremia with sodium level of 118 mEq/L, and she becomes more lethargic. What are some causes of hyponatremia and which is the most likely in this case?*
   Various causes of hyponatremia are listed below.
   - Hypovolemic hyponatremia: sodium and free water loss with inappropriately hypotonic fluid replacement
     - Cerebral salt-wasting syndrome: traumatic brain injury, aneurysmal subarachnoid hemorrhage, subdural hematoma, and postcraniotomy—the most likely cause in this case
     - Gastrointestinal losses due to vomiting or diarrhea
     - Excessive sweating
     - Third spacing of fluids (peritonitis, pancreatitis, burns)
     - Acute or chronic renal insufficiency
     - Prolonged exercise in a hot environment
   - Euvolemic hyponatremia: normal sodium stores and total body excess free water
     - Psychogenic polydipsia
     - Hypotonic IV or irrigation fluids postoperatively
   - Hypervolemic hyponatremia: inappropriate increase in sodium stores
     - Hepatic cirrhosis
     - Congestive heart failure
     - Nephrotic syndrome
     - Hypothyroidism
     - Cortisol deficiency
     - Syndrome of inappropriate antidiuretic hormone (SIADH)[11]
     - Medications: acetazolamide, thiazide diuretics, angiotensin-converting enzyme inhibitors, carbamazepine, gabapentin, haloperidol, heparin, ketorolac, loop diuretics, nimodipine, opiates, proton pump inhibitors, and selective serotonin reuptake inhibitors[11–13]
7. *How would you manage this problem?*
   - Ensure that the patient is stabilized first: secure airway, breathing, circulation (ABCs of trauma), place on oxygen transfer to intensive care unit (ICU), obtain IV access with large bore IV, and place a Foley catheter
   - Obtain following laboratory studies to determine the type of hyponatremia:

## Answers (*continued*)

   - Serum and urine sodium and osmolalities, and urine specific gravity
   - Serum electrolytes
   - Thyroid function studies
   - Adrenal function studies
- In this case, the most likely cause is hypovolemic hyponatremia possibly due to the subdural hematoma.
- If the symptoms are mild to moderate, one may treat with isotonic saline.
- Monitoring of serum sodium levels needs to be done frequently to ensure that the serum sodium does not increase more than 0.5 mEq/L/hour or 12 mEq/L/day.
- Hypertonic saline (3%) may be used as an alternative if normal saline alone is not adequate enough to correct the problem. When using this solution, care should be taken to administer it slowly (20 to 50 cc/hour drip) and frequent checks on serum sodium are required (every 2–4 hours).
- Evacuation of the subdural hematoma may resolve the hyponatremia (it did in this case).[12,13]

8. *What is your course of action now?*
   - Urgent ICU admission
   - Laboratory studies: complete blood count, electrolytes, coagulation profile, type and screen, blood, urine, sputum cultures, and culture discharge from wound
   - Obtain imaging studies: CT and MRI with and without contrast
   - May elect to wait until the cultures are sent intraoperatively before starting any broad-spectrum antibiotics
   - Antibiotics suggested:
     - Vancomycin 1 g IV every 12 hours, cefepime 2 g IV every 12 hours, and metronidazole 500 mg IV every 8 hours
     - Gentamycin 600 mg IV every 8 hours may be used instead of cefepime, if the patient is allergic to penicillin.
     - Antibiotic changes are performed as culture and sensitivity results are obtained.
9. *A follow-up CT scan and subsequently an MRI scan are obtained (▶* **Fig. 55.2***). Interpret the studies and provide a differential diagnosis.*
   - CT scan with contrast shows some reaccumulation of fluid bilaterally. The fluid collection on the right side is partly hyperdense or enhancing, which is suggestive of acute blood or possible empyema with peripheral enhancement.
   - On MRI, the subdural fluid is consistent with blood of different ages and it is seen bilaterally. On the right side, there is some enhancement surrounding the subdural fluid which is possibly suggestive of infection.
   - The right subdural fluid appears to be iso- to hyperintense on Tl-weighted and hyperintense on T2-weighted or fluid-attenuated inversion recovery (FLAIR) MRIs. If it is blood, it is likely subacute to chronic (3 weeks old).
   - The differential diagnosis includes:
     - Reaccumulation of subdural hematoma
     - Subdural empyema
     - Subdural hygroma
10. *What is your management plan?*
    - Urgent surgical evacuation via burr hole reexploration on the right side[14–16]
    - The exposure should be prepared in the event that a craniotomy is needed.
    - Subdural fluid needs to be sent for culture as soon as possible, after which the antibiotics are initiated.
    - Incision and sharp debridement of the posterior scalp wound is needed with irrigation and antibiotics and saline solution.
    - A drain can be left in the subgaleal space.
    - Wound closure can be done in a single layer with 2–0 Prolene (Ethicon, Somerville, NJ) vertical mattress interrupted sutures to promote better healing of infected or granulating tissues.

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# Case 56 Epidural Hematoma

*Abdulrazag Ajlan and Judith Marcoux*

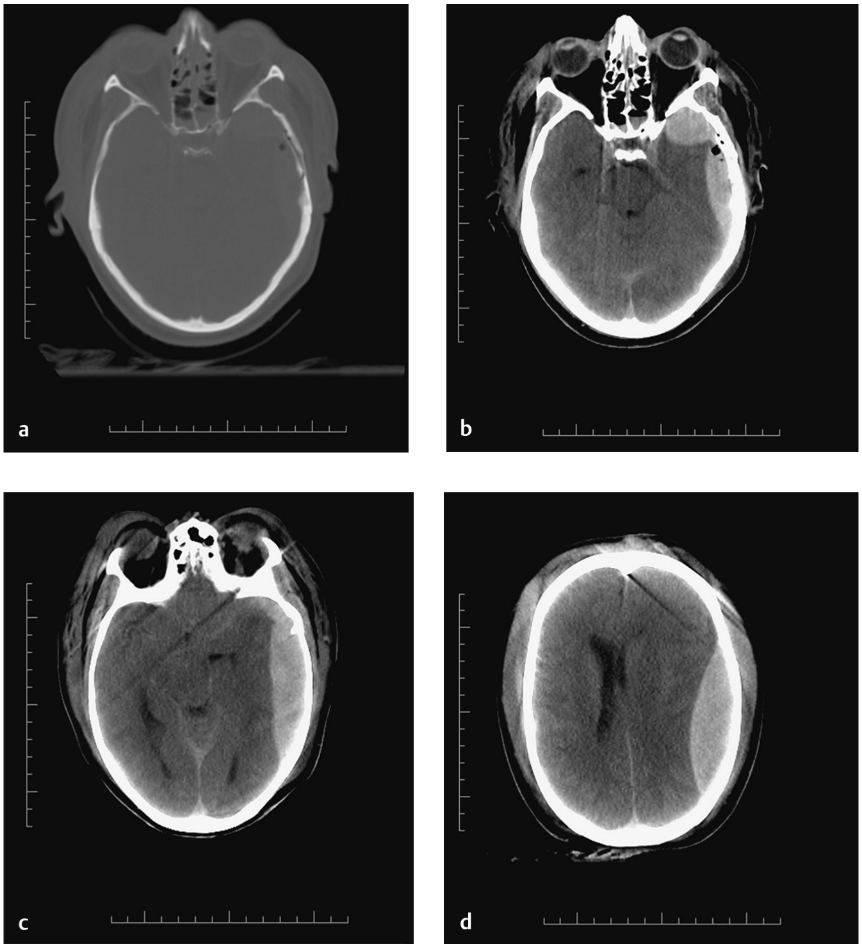

**Fig. 56.1** CT scan of the head, (**a**) bone window where a left temporal bone fracture is seen. (**b**), (**c**), and (**d**) are brain windows showing a large epidural hematoma with significant mass effect and uncal herniation with brainstem compression and displacement (**c**) and significant midline shift (**d**).

## Clinical Presentation

- A 42-year-old woman fell from the third floor of a building. She presents with a Glasgow Coma Scale (GCS) score of 7.
- The patient is hemodynamically stable and she is quickly intubated.
- Her pupils are both reactive, but asymmetrical.

## Questions

1. Describe what you see on the CT scan (▶**Fig. 56.1**).
2. What is the pathophysiology of this intracranial bleed?
3. What is your initial management?
4. What are the criteria for surgical evacuation?
5. What is the prognosis?

## Answers

1. *Describe what you see on the CT scan (▶* **Fig. 56.1***).*
   - There is a linear nondisplaced temporal bone fracture with underlying pneumocephalus.
   - There are also hyperdense lesions in the temporal and temporoparietal areas on the same side of the fracture. The lesions are biconvex (lens shape) and are compatible with an epidural hematoma (EDH).
   - The lesions are causing a midline shift of 0.7 cm and a right-sided uncal herniation.
2. *What is the pathophysiology of this intracranial bleed?*
   - EDHs are usually located in the temporoparietal areas; posterior fossa hematomas represent 5% of the EDH.
   - The source is usually a meningeal artery, which is most commonly the middle meningeal artery, but sometimes this can be a bleeding vein or an underlying sinus.
   - The bleeding in the epidural space will strip the dura from the skull causing mass effect and raising intracranial pressure.
   - Approximately 85% of cases will be associated with skull fracture.
   - Twenty percent of patients will present in a comatose state.[1] Other presentations include localized neurologic findings and confusion. The classical presentation of transient improvement followed by sudden deterioration occurs in 47% of admissions, which is called the lucid interval.[2]
3. *What is your initial management?*
   - Once the airway is secured and the patient is hemodynamically stable, she needs to undergo emergency evacuation for the hematoma.
   - The patient should also be loaded with an antiepileptic drug.
   - Infusing mannitol and hyperventilation may be done as temporary measures before the evacuation.
   - This is a surgical emergency because there is a symptomatically significant mass > 1 cm with midline shift > 0.5 cm.
4. *What are the criteria for surgical evacuation?*
   - An EDH with volume > 30 $cm^3$ regardless of the GCS score.[2]
   - A patient with a GCS score > 8 and a hematoma < 30 $cm^3$, with < 15 mm thickness, < 0.5 cm midline shift, and without focal deficit can be managed by close observation with serial scans and placement in a monitored neurosurgical unit.[2]
5. *What is the prognosis?*
   - Outcome depends on many factors, for example, GCS score, CT scan findings, age, timing of surgery, and the presence of other lesions.
   - The overall mortality for patients who underwent surgical evacuation is 10%.[2] The faster the evacuation is done, the better will be the outcome because the neurologic status prior to surgery is the main determinant of outcome.
   - EDH evacuation is considered one of the most “cost effective” surgical procedure of all in terms of quality of life.

## Suggested Readings

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# Case 57 Traumatic Acute Subdural Hematoma

*Abdulrazag Ajlan and Judith Marcoux*

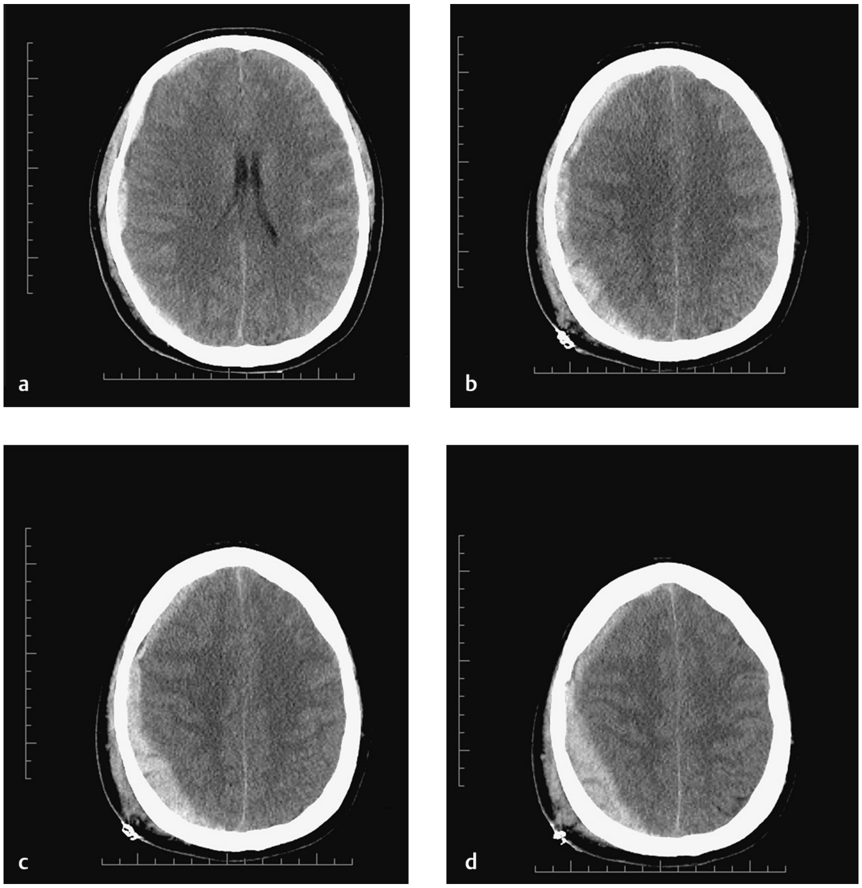

**Fig. 57.1** CT scan of the head, brain windows showing a right-side subdural hematoma with associated midline shift > 5 mm (**a**) and brain compression (**b–d**).

## Clinical Presentation

- A 39-year-old man is involved in an all-terrain vehicle accident; he had no helmet on.
- His Glasgow Coma Scale (GCS) score on arrival in the emergency room was 7 (eyes 1, verbal 1, motor 5).
- The patient is hemodynamically stable and is quickly intubated.

## Questions

1. Describe what you see on the CT scan (▶**Fig. 57.1**).
2. What is the pathophysiology of this intracranial bleed?
3. What is your management?
4. What are the criteria for surgical evacuation for acute subdural hematomas (SDHs)?
5. What is the prognosis?
6. What other types of traumatic intracranial hemorrhage can you see?
7. What are the indications for surgical evacuation in the different types of intracranial hemorrhage?
8. What are the indications for antiepileptic medication?

## Answers

1. *Describe what you see on the CT scan (▶ **Fig. 57.1**).* There is an acute SDH over the right hemisphere that is causing mass effect, compression of the brain, and a midline shift.
2. *What is the pathophysiology of this intracranial bleed?*
   - Acute SDHs usually occur due to torn bridging veins at the surface of the brain. Damaged cortical arteries could also produce SDHs.
   - A significant degree of impact is usually required to produce a SDH.
3. *What is your management?*
   - This patient is comatose; he has a mass lesion seen on the CT scan and significant midline shift associated with it (> 5 mm).
   - After ensuring that there is no life-threatening injury, this patient should be taken to the operating room for quick decompression via a large frontotemporoparietal craniectomy or craniotomy.
4. *What are the criteria for surgical evacuation for acute subdural hematomas (SDHs)?*
   - Indications for surgery are as follows:[1]
     - An acute SDH with a thickness > 10 mm or a midline shift > 5 mm on CT scan should be surgically evacuated, regardless of the patient's GCS score.
     - All patients with an acute SDH in a coma (GCS less than 9) should have an intracranial pressure (ICP) monitor.
     - A patient with a GCS score less than 9, an acute SDH < 10 mm, and a midline shift < 5 mm should undergo surgical evacuation of the lesion if one of the following is present:
       - Decrease in the GCS score by 2 or more points
       - Asymmetric or fixed and dilated pupils
       - Elevated ICP of more than 20 mm Hg
5. *What is the prognosis?*
   - The prognosis will depend on the initial GCS score, the age, and the associated brain damage (with subsequent intracranial hypertension) of the patient.
   - Early surgery may improve the outcome.
6. *What other types of traumatic intracranial hemorrhage can you see?*
   - The other types of traumatic intracranial bleeding, apart from acute SDH and acute epidural hemorrhage, include:
     - Intraventricular hemorrhage
     - Subarachnoid hemorrhage
     - Intracerebral contusions which are of three subtypes:
       - Coup contusions (occur at the site of the impact)
       - Contrecoup contusions (occur at sites remote from the impact due to movements of the brain inside of the skull; they are most commonly found in the frontal and temporal poles and the subfrontal area.)
       - Gliding contusions (occur due to herniation or tissue tear)
     - Intracerebral hematomas (greater content of blood compared with contusions); usually caused by shearing of small vessels, but could also be caused by traumatic aneurysms.
7. *What are the indications for surgical evacuation in the different types of intracranial hemorrhage?*
   - An intraventricular hemorrhage can cause hydrocephalus requiring cerebrospinal fluid drainage. Subarachnoid hemorrhage does not require surgical evacuation; however, if it is abundant enough, it may cause vasospasm.
   - Contusions could be small and limited to the cortical layer, but they could also cause significant mass effect. Furthermore, contusions have the tendency to increase in size over the first 24 to 48 hours (and sometimes even longer) following a trauma. The prognosis following traumatic contusions will depend on the location, number, and the size of the contusions. Contusions may be a cause of posttraumatic seizure. Due to their location, they can also play a role in cognitive and behavioral deficit following a traumatic brain injury (TBI).
   - Patients with mass effect, worsening neurologic exam, or refractory high ICP should undergo surgical evacuation. Patients with a GCS score of 6 to 8 with frontal or temporal contusions > 20 $cm^3$ with a midline shift ≥ 5 mm or compression of the basal cisterns should have surgery. Any contusion with a volume > 50 $cm^3$ should be evacuated.[2] Any lesion of the posterior fossa associated with mass effect or neurologic dysfunction should be evacuated.[3]
8. *What are the indications for antiepileptic medication?*
   - Prophylaxis of late posttraumatic seizure is not recommended.
   - Prophylaxis of early posttraumatic seizure (within 7 days) is recommended, but has not been shown to change the overall outcome.[4]

## Suggested Readings

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# Case 58 New Trends in Neurotrauma Monitoring

*Judith Marcoux and Abdulrazag Ajlan*

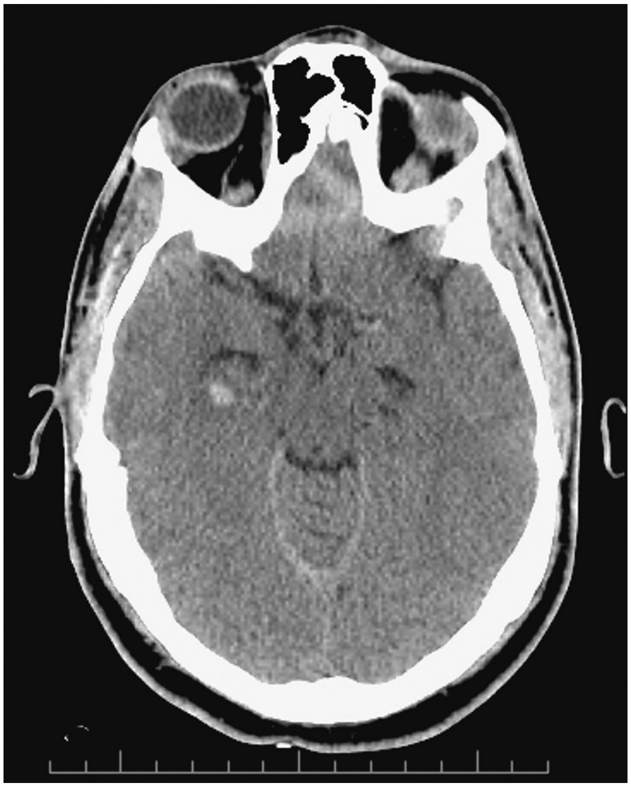

**Fig. 58.1** CT scan of the head showing a very small intraparenchymal hematoma in the right hippocampal area.

## Clinical Presentation

- A 20-year-old man is involved in a high-speed motor vehicle accident. He was the driver and was wearing his seatbelt.
- He is hemodynamically stable and his Glasgow Coma Scale (GCS) score before intubation is 8.
- A CT scan of his head is obtained and shown in ▶**Fig. 58.1**.
- His GCS remains 8 without sedation and he has an intraventricular drain placed to monitor his intracranial pressure (ICP). The opening pressure is 17 mm Hg.
- He is kept normothermic, normocapnic, and with a cerebral perfusion pressure (CPP) above 70 mm Hg.
- Over the course of the next 3 days, his ICP rises and he needs heavier sedation, cerebrospinal fluid (CSF) drainage, and hyperosmolar therapy.
- A repeat CT scan shows diffuse cerebral edema with effacement of the subarachnoid spaces.
- Transcranial Doppler (TCD) measurements reveal high velocities in both carotid arteries as well as the middle cerebral arteries.

## Questions

1. What other tools can help you optimize his ICP management?
2. What is the role of continuous electroencephalography (EEG)?
3. What is the role of microdialysis?

## Answers

1. *What other tools can help you optimize his ICP management?*
   - Jugular venous oxygen saturation ($SjvO_2$):
     – The oxygen saturation in the brain draining veins will depend on the cerebral blood flow (CBF), the metabolic rate of oxygen consumption, as well as the hemoglobin concentration. Under normal circumstances, the saturation will be between 55 and 70%.
     – However, if the cerebral tissue does not receive enough oxygen for its metabolic demands (due to low flow or low transportation of oxygen due to low hemoglobin), the $SjvO_2$ will drop, reflecting an increased extraction of oxygen from the brain.
     – On the other hand, if the metabolic rate of oxygen in the brain is decreased, or the blood flow is high, the $SjvO_2$ will increase. Therefore, a supranormal value of $SjvO_2$ could mean a state of hyperemia. But it could also reflect an impaired extraction capacity of the brain, which can be seen following a traumatic brain injury (TBI). $SjvO_2$ will detect global changes, but may miss regional abnormalities.
     – $SjvO_2$ clinical use is not simple; however, it does have some applications. Hyperventilation is very effective in lowering the ICP, but it does so by reducing the CBF. Because brain tissue is very sensitive to ischemia following an acute TBI, one must be careful not to overuse hyperventilation. One way to monitor the effect of hyperventilation on cerebral perfusion is by measuring the $SjvO_2$.
     – If the hyperventilation is excessive, the $SjvO_2$ will drop and the ventilator settings can be quickly adjusted (level III recommendation by the Brain Trauma Foundation).[1]
     – Low $SjvO_2$ has also been linked with poor outcome.[2]
   - Brain tissue partial pressure of oxygen ($PO_2$):
     – Human brain is dependent on a good oxygenation for its metabolism and a decrease in tissue oxygenation will increase the risk of ischemic cell damage, especially following a TBI. Brain tissue oxygenation is dependent on CPP, but local hypoxia can happen despite normal CPP, ICP, and mean arterial pressure.
     – Monitoring brain tissue $PO_2$ can help detect an area at risk of ongoing damage. Recent evidences seem to show that aggressively treating the $PO_2$ will improve the outcome.[3] One must bear in mind, however, that it reflects the metabolic state of a small area of the brain only, other areas may differ significantly.
   - CBF
     – Monitoring the CBF when the ICP is elevated may help differentiate an ischemic state from a hyperemic state and dramatically change the therapeutic approach to the TBI patient.[4]
     – There are different ways to measure CBF; the most commonly used being the TCD, which is a bedside noninvasive method, but is usually noncontinuous.
     – In the current case, the increased velocities of TCD reflect hyperemia. Lowering the CPP to 60 mm Hg helped control the current patient's ICP.
2. *What is the role of continuous electroencephalography (EEG)?*
   - Posttraumatic seizures complicate a sizable percentage of TBIs. The risk will increase with the severity of the trauma.
   - In patients with moderate and severe TBI, 22.3% had electrical seizures on continuous EEG monitoring. More than half of which had no clinical manifestation of their seizures.[4]
   - In critically ill comatose patients, the duration of nonconvulsive status epilepticus and the delay in diagnosis was associated with increased mortality.[5]
   - Continuous EEG monitoring may help diagnose and treat nonconvulsive seizures, and thus prevent secondary injury to the brain. Furthermore, continuous EEG is of primary importance in the titration of barbiturate-induced coma to treat raised ICP.
3. *What is the role of microdialysis?*
   - Microdialysis gives a unique insight into the brain's metabolism by measuring tissue biochemistry.
   - It can give a warning regarding impending hypoxia/ischemia or the occurrence of secondary damage. But its measure is local and there is a great variability in the range of the values collected. Its use is still limited to research. However, a growing number of clinicians are using it. In 2004, a consensus[6] was made for its use and some of the major points are as follows:
     – The use of microdialysis should be reserved for patients with a severe TBI requiring ICP monitoring.
     – The probe should be placed in the right frontal area in patients with diffuse lesions, or placed in the pericontusional area if there is a focal lesion.
     – The best marker for secondary damage is the lactate:pyruvate ratio; glucose, glutamate, and glycerol can also be used as markers of ischemia.

## Suggested Readings

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# Case 59 Intracranial Pressure Management

*Abdulrazag Ajlan and Judith Marcoux*

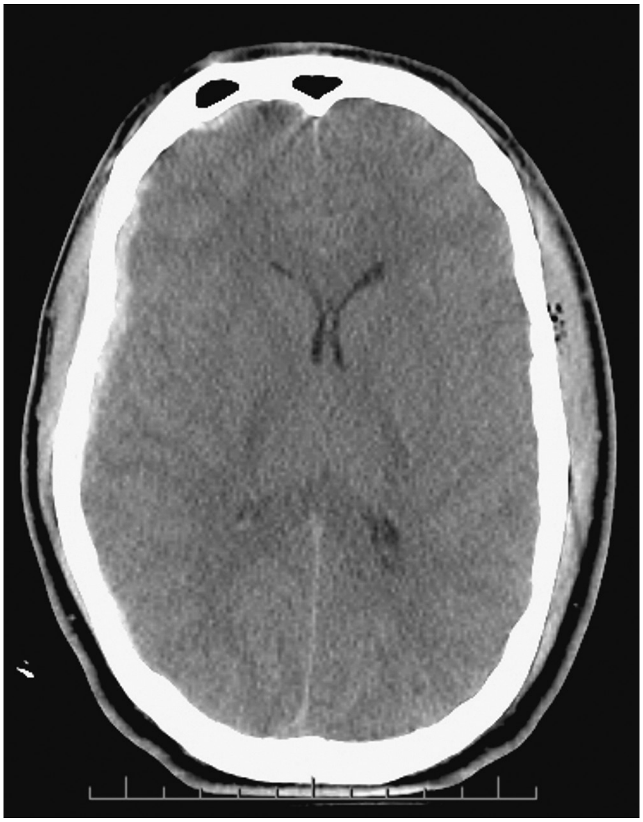

**Fig. 59.1** A brain CT scan showing a small right subdural hematoma with a midline shift of 4 mm. There is diffuse brain swelling and small ventricles.

## Clinical Presentation

- A 26-year-old male pedestrian is hit by a car.
- Initial Glasgow Coma Scale (GCS) score is 6 after resuscitation.
- A CT scan is obtained and it shows a small right acute subdural hematoma (SDH) with a 4 mm midline shift (▶**Fig. 59.1**).
- An intraventricular drain is placed to monitor the intracranial pressure (ICP).

## Questions

1. What is the pathophysiology of raised ICP?
2. How can we monitor the ICP?
3. What are the indications for ICP monitoring?
4. How can we manage elevated ICP (describe first-, second-, and third-tier measures)?

## Answers

1. *What is the pathophysiology of raised ICP?*
   - The Monro–Kellie hypothesis states that the cranial compartment is incompressible, and the volume inside the cranium is a fixed volume.
   - The cranium and its constituents (blood, cerebrospinal fluid [CSF], and brain tissue) create a state of volume equilibrium, such that any increase in volume of one of the cranial constituents must be compensated by a decrease in volume of another.[1,2]
   - Small increases in brain volume do not lead to immediate rise in ICP because of compensatory measures such as displacement of CSF into the spinal canal, stretching of the falx cerebri and the tentorium, and decrease in venous blood volume. However, once the ICP goes beyond the compensatory phase, small increases in brain volume (or any other constituent) can lead to marked elevations in the ICP (▸**Fig. 59.2**).
   - Once the ICP starts to elevate, it affects the brain in two major ways:
     - High ICP leads to decreased cerebral blood flow (CBF) as can be observed by the following equations:
       - ○ CPP = MAP − ICP
       - ○ CBF = CPP/CVR

     where, CPP is cerebral perfusion pressure, MAP is mean arterial pressure, and CVR is cerebral vascular resistance.
     - Any increase in the ICP will decrease the CPP. In the normal physiologic state, the CBF will remain constant because the CVR will vary to compensate, this is called autoregulation. This compensation is impaired in the extreme of pressure or in abnormal states (e.g., brain trauma).
   - Once the ICP is high and uncompensated, the brain tissue will start shifting and herniating through dural openings. This herniation will cause brain tissue compression, disturbance in blood flow, damage to the vasculature, as well as further increase in the ICP. There are different types of herniation:
     - Uncal herniation is caused by a mass in the middle fossa that displaces the uncus between the midbrain and tentorial edge. This causes compression on the descending tract and the reticular formation in the brainstem causing a decrease in the level of consciousness and contralateral hemiparesis. The ipsilateral oculomotor nerve is also affected causing ptosis and mydriasis (▸**Fig. 59.3** and ▸**Fig. 59.4**).
     - Sometimes the midbrain is squeezed against the contralateral tentorial edge (Kernohan's notch) causing ipsilateral hemiparesis (▸**Fig. 59.3** and ▸**Fig. 59.4**). The posterior cerebral artery will be compressed causing ischemia.
     - With more severe herniation, the basilar artery will be stretched causing tearing of the brains-

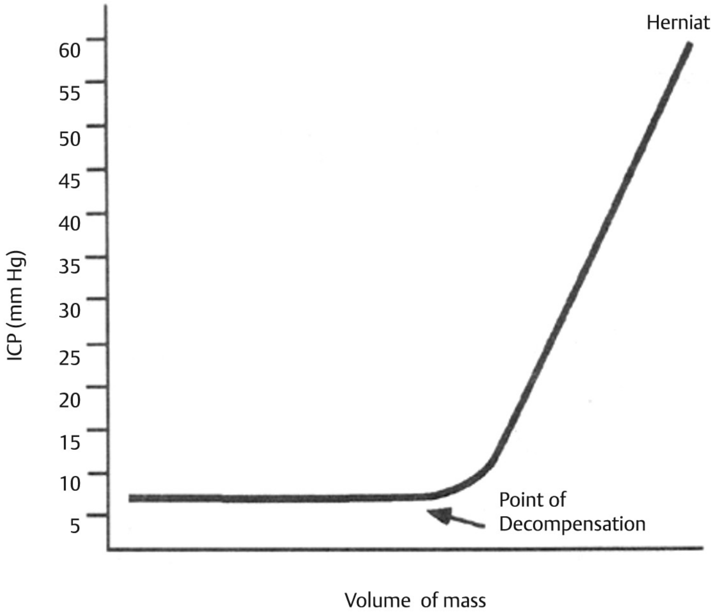

**Fig. 59.2** The brain can compensate within a limited range of pressure. After the compensatory mechanisms fail, the intracranial pressure will increase dramatically.

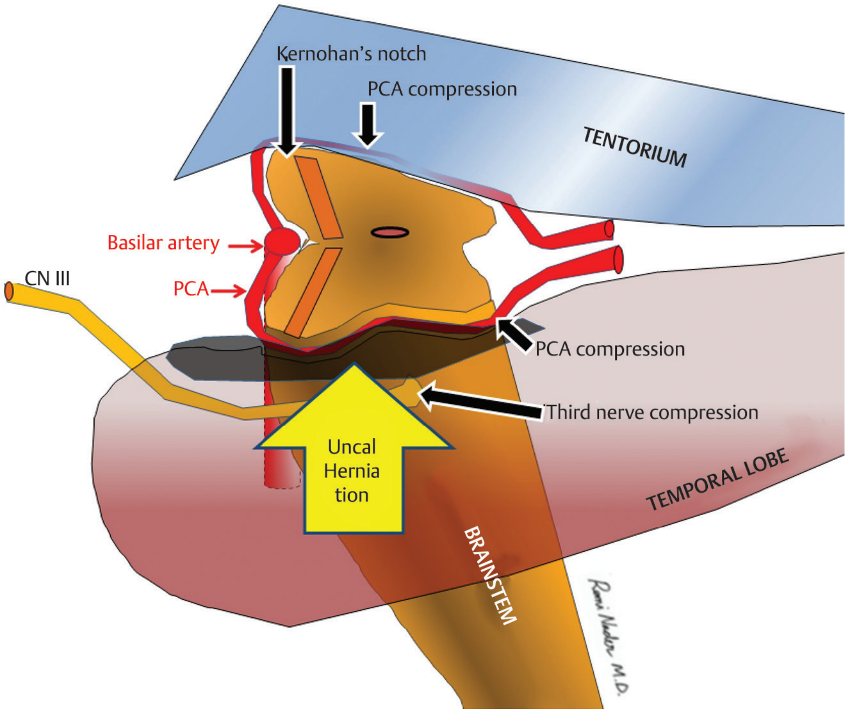
Kernohan's notch
PCA compression
TENTORIUM
Basilar artery
CN III
PCA
PCA compression
Third nerve compression
Uncal Hernia tion
TEMPORAL LOBE
BRAINSTEM

**Fig. 59.3** Herniation of the uncus is causing compression of the brainstem, posterior cerebral artery (PCA), and oculomotor nerve (CN III). Compression will normally cause ipsilateral pupil dilatation and contralateral hemiparesis.
If the brainstem is shifted toward the other side, structures on that side may be compressed, giving instead a contralateral pupil dilatation and an ipsilateral hemiparesis.

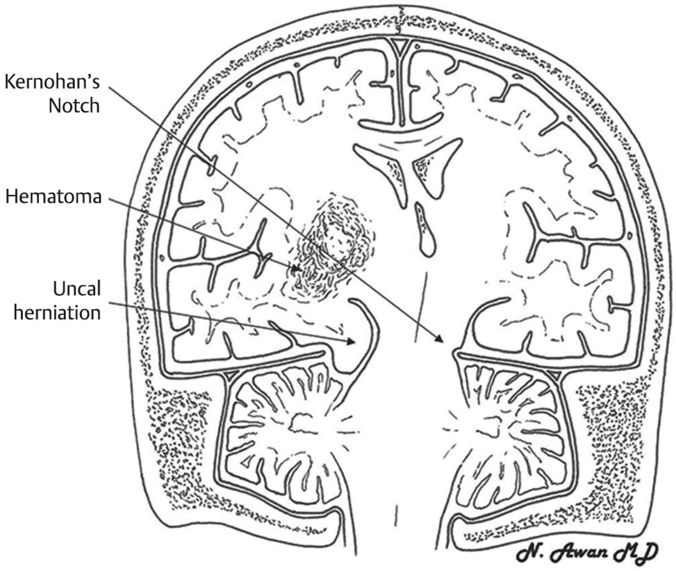
Kernohan's Notch
Hematoma
Uncal herniation
N. Awan M.D

**Fig. 59.4** Artist's rendering of uncal herniation.

## Answers (*continued*)

tem perforator vessels, brainstem infarction, and bleeding (Duret's hemorrhages).
  - The Cushing triad occurs due to high ICP and herniation. This triad includes hypertension, bradycardia, and respiratory irregularities.

2. *How can we monitor the ICP?*
   - One of the most reliable methods is the clinical picture of the patient. If the patient is awake enough to be followed clinically, this can be used as an objective parameter indicating that he is maintaining his perfusion and that his ICP is compensated. Once the clinical exam is lost for any reason (e.g., decreased level of consciousness or intoxication), another parameter should be used.
   - Several methods have been developed for measuring the ICP. The classical methods include:
     - Intraventricular catheter placement (ventriculostomy)
     - Intraparenchymal monitoring
     - Epidural devices
     - Subdural/subarachnoid devices (bolts)

## Answers (*continued*)

- The ventricular catheter is the gold standard given that it is the most accurate method. It is also therapeutic as well as diagnostic.

3. *What are the indications for ICP monitoring?*
   - The guidelines were revised by the Brain Trauma Foundation in 2007[3] and include:
     - Patient with severe injury (GCS ≤ 8) after resuscitation and with an abnormal CT scan (hematoma, contusion, edema)
     - Patient with severe injury (GCS ≤ 8) and normal CT scan with two or more of the following:
       - ○ Age above 40 years
       - ○ Unilateral or bilateral motor posturing
       - ○ Systolic blood pressure < 90 mm Hg
   - ICP monitoring is not routinely indicated in mild or moderate brain trauma cases. However, the treating physician can choose to monitor ICP in certain conscious patients with abnormal CT scan findings.[3]
4. *How can we manage elevated ICP (describe first-, second-, and third-tier measures)?*
   - Brain injury from trauma results from:
     - Primary injury from the first impact. This type of damage has no treatment other than prevention
     - Secondary injury, which can be due to:
       - ○ Intracranial causes such as expanding hematomas, contusions, or diffuse edema that lead to rise in ICP
       - ○ Systemic causes such as hypotension, hypoxia, and pyrexia that lead to mismatch in the metabolic demand and the blood flow.
   - Therefore, the treatment starts at the scene by preventing hypotension and hypoxia with adequate resuscitation and airway management, followed by a rapid transfer to a trauma center.
   - Once the CT scan is done and a surgical mass lesion is ruled out, an ICP monitor is inserted if indicated.
   - The goal of treatment is to keep an adequate blood flow which will match the metabolic demand of the injured brain. To reach this goal, the ICP should be kept below 20 mm Hg,[4] and the CPP should be between 50 and 70 mm Hg.[5] First-, second-, and third-tier therapies have been devised for ICP treatment.
   - First-tier therapies:
     - Elevation of the head to 30 degrees to enhance the venous outflow
     - Keeping the neck straight and the venous outflow patent
     - Ventilation to normocarbia ($pCO_2$ = 35–40 mm Hg) to prevent cerebral vasodilatation. Hyperventilation can reduce ICP by causing cerebral vasoconstriction. On the other hand, this decrease in CBF can cause ischemia. Therefore, the use of hyperventilation as a prophylactic measure with $pCO_2 < 25$ mm Hg is not recommended, especially in the early period after trauma. Hyperventilation is only recommended as a temporary measure in the case of high ICP until other treatments are started or with other monitoring tools for the CBF (see **Case 58**, New Trends in Neurotrauma Monitoring).
     - Avoid hyperthermia because it may lead to an increase in the metabolic rate. Acetaminophen and a cooling blanket can be used to achieve this goal.
   - Second-tier therapies:
     - Agitation and pain are two common causes of elevated ICP. Thus, sedation and pain control present an effective treatment modality.
     - Hyperosmolar treatment is used to decrease brain edema and subsequently the ICP. The hyperosmolar treatment will also help in cerebral perfusion.[6] Mannitol and hypertonic saline can be used as osmotic therapy agents.
     - Neuromuscular paralysis for maximal muscle relaxation and decreased muscle tone
     - CSF drainage from the intraventricular drain
   - Third-tier therapies:
     - Decompressive craniectomy can be very effective in reducing the ICP. However, it is not clear if this measure improves the outcome. There are no clear guidelines about indications and timing. The only randomized clinical trial was done in a pediatric group. It shows a better outcome, but this did not reach statistical significance.[7]
     - Barbiturate coma: high-dose barbiturate therapy can result in control of ICP when all other medical and surgical treatments fail. This effect is attributed mainly to the decrease in the cellular metabolic rate and subsequent decrease in CBF. The main side effects are hemodynamic instability and lowered immunity. It has shown no clear benefit in improving outcome and the prophylactic use in the treatment of ICP is not recommended.[8]

## Suggested Readings

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# Case 60 Gunshot Wound Injury to the Head

*Faisal Abdulhamid Farrash*

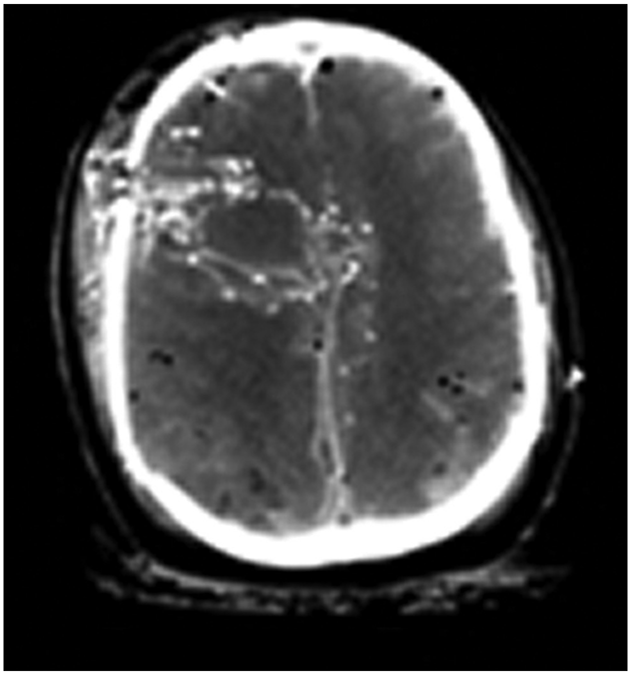

**Fig. 60.1** Axial CT scan without contrast of patient described herein.

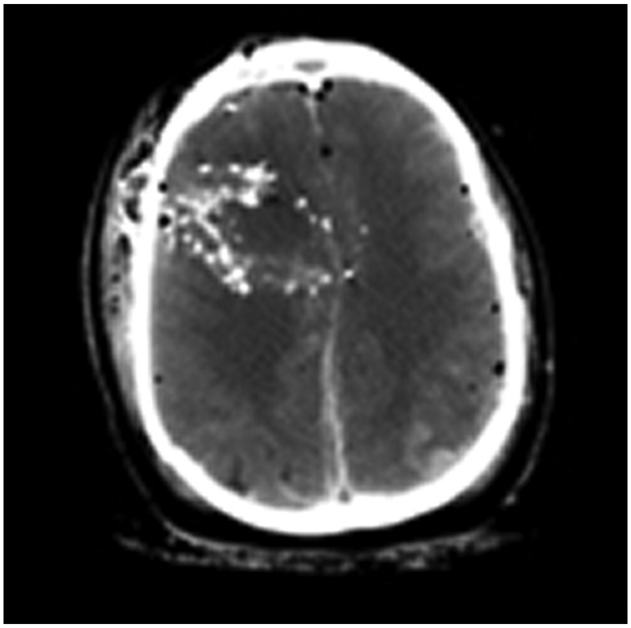

**Fig. 60.2** Axial CT scan without contrast of patient described herein.

## Clinical Presentation

- A 50-year-old man is rushed to the ER after being involved in an altercation.
- Physical examination reveals that he is unconscious and actively bleeding from a right forehead wound, he is breathing spontaneously hypotensive and tachycardic with a Glasgow Coma Scale (GCS) score of 8. The right pupil is 6 mm with slow reaction to light. The left pupil is round, regular, and reactive to light.
- Carefully inspecting the wound, you find burn marks and black powdered material around the periphery of the wound as well as some brain matter and bone debris.

## Questions

1. What is the most likely diagnosis? Briefly describe its epidemiology.
2. What are your initial steps in management?
3. What are the questions that come to your mind while taking a history from this patient's family and witnesses present at the scene?
4. What kind of radiological studies would you order or consider ordering on this patient and why?
5. Interpret the images in ▶**Fig. 60.1**, ▶**Fig. 60.2**, and ▶**Fig. 60.3**. Highlight the important findings and indicate what you look for on those images.
6. Should you give prophylactic antibiotics? What are the factors that may raise the incidence of infection in such cases?
7. Describe the mechanism of damage in gunshot injuries to the brain.
8. What are the indications for surgery and why?
9. What type of intervention would you offer this patient and why? How would you plan it?
10. What is the expected outcome of such patients?
11. What factors related to weapons and ammunition might affect the severity of the injury?
12. In penetrating injury to the head due to a stab, when do you attempt to remove the offending object?

## ■ Answers

1. *What is the most likely diagnosis? Briefly describe its epidemiology*.
    - Given the patient's history, the most likely diagnosis is penetrating head injury (i.e., gunshot wound to the head).
    - This type of injury usually causes a depressed or comminuted open skull fracture with intracranial hemorrhage and penetrating head injury.
    - The injury may also be accompanied by black or dark skin discoloration around the entry wound and this is mainly seen when the gun has been held in very close proximity to the skin.[1] Gunpowder marks may also be seen in rifle injuries where the weapon is held at a short distance.[2]
    - Differential diagnosis includes any type of penetrating head injury from a blast or high-energy shrapnel from a nearby explosion.
    - Gunshot wounds are high-mortality injuries with 71% of civilian casualties immediately dying following the injury.[3]
        - Mortality rates reach 15% in young healthy patients below 40 years and 70% in patients above 60 years of age who make it to the hospital.[4]
        - These are more common in males than females[5] which could be attributed to the larger numbers of male soldiers world-wide as well as the higher number of males being involved in suicidal and homicidal injuries involving firearms compared to females.
        - Attributable factors are usually homicidal crimes or military conflicts in injuries occurring between the ages 20 to 30 years.
        - Attributable factors are usually suicidal attempts in subjects of age 60 years and above. This group fares worse than the former age group.
        - Location of injury is typically along the right temple in suicidal attempts compared to other types of firearm injuries[6] (see ▶**Table 60.1** for details of distribution).
2. *What are your initial steps in management?*
    - The initial steps in management of this patient must be performed in the following order:
        - Securing the patient's Airway (do not forget C-spine immobilization in unconscious patients).
        - Maintaining the patient's Breathing by a face-mask, bagging, or by intubation and ventilatory support. This helps in controlling his breathing parameters and also presents a tool to reduce an elevated intracranial pressure (ICP).
        - Circulation maintenance: based on the described clinical scenario, the patient is in hemodynamic shock and needs rapid treatment. This includes arresting the current source of bleeding and initiating replacement of deficient intravascular volume.
            - ○ One should immediately place two large-bore IV cannulae or a central line to start fluid replacement (the central line can also aid in monitoring the central venous pressure).
            - ○ A type and cross match for four units of blood should be obtained.
            - ○ Foley's catheter is placed that helps in estimating tissue perfusion via strict input and output charting.
            - ○ The use of inotropic support maybe needed to enhance the patient's blood pressure and cardiac contractility.
        - Disability (D) evaluation and Exposure (E) of the entire body for screening of additional injuries should also be performed and are included in the primary survey based of the Advanced Trauma Life Support (ATLS) guidelines, which should be completed in a fast and efficient manner.
    - Note: one should not forget seizure prophylaxis. Penetrating head injuries are highly epileptogenic in the acute setting with a risk > 50% of seizures and a high risk of delayed seizures as well (39%).[8] Seizures control in the acute setting does not affect the possibility of having delayed seizures in the future.[9] Acute seizure prophylaxis for 7 days plus a loading dose is recommended in the treatment guidelines for management of penetrating brain injuries.[10]
3. *What are the questions that come to your mind while taking a history from this patient's family and witnesses present at the scene?*
    - Questions that must be asked during history taking include:
        - Were there any other bullet injuries elsewhere?
        - Was there involvement in any fight or any violent or criminal activity?
        - Was the injury acquired in the battlefield? This helps in determining the type or category of the weapon used.
        - Is there any history of mental or psychiatric disorders? Suicidal attempts are associated with a worse outcome.[3]
        - What was the type and caliber of the weapon used?
        - What was the distance of the weapon from the victim?
        - How many bullet injuries are there? Multiple gunshot injuries to the head are typically incompatible with life.
        - Was there any explosion or blast at the scene?
        - What was the direction of the bullet or shrapnel?
        - Where was the assailant standing?
    - All of these questions will help in determining the magnitude and trajectory of the bullet. It would also help in formulating a proper, individualized treatment plan.

## Answers (*continued*)

4. *What kind of radiological studies would you order or consider ordering on this patient and why?*
   - CT scan of the brain is mandatory in such cases after initial stabilization. 3D reconstruction is also helpful in measuring the amount of bone deficits if planning a potential surgical reconstruction.
   - Ordering other types of radiological investigations depends on the patient's clinical status and the need to rule out other injuries.
     - For example, CT angiography (CTA) or cerebral angiogram to evaluate for fistulas, aneurysms[11] or psudoaneurysms[12] and rarely to confirm brain death.
     - These studies may be needed especially if the bullet path crosses major central nervous system (CNS) vascular landmarks.
     - CT with contrast may be obtained to rule out brain abscess or empyema development. This typically does not develop at first but rather over a longer time-frame.
   - Ordering other radiological exams need to be tailored according to individual patient presentation. Obtaining unnecessary exams may waste precious time that can be used in saving this patient's life or vital functions. The viability of the brain and spinal cord tissue compared to other types of tissue must also be taken into consideration in such injuries.
   - As a general rule, MRI should not be obtained in cases of gunshot wound to the head, as residual metallic bullet fragments may be displaced by the magnetic field and can cause further injury to the surrounding brain.
5. *Interpret the images in* ▶**Fig. 60.1**, ▶**Fig. 60.2**, *and* ▶**Fig. 60.3**. *Highlight the important findings and indicate what you look for on those images.*
   - The images show a CT scan (▶**Fig. 60.1** and ▶**Fig. 60.2**) of the brain with bone windows revealing the entry site of the bullet, as well as bone fragments and debris dispersed inside the brain parenchyma surrounding the created bullet cavity. There are multiple scattered pockets of pneumocephalus as well as diffuse traumatic subarachnoid hemorrhage (SAH).
   - The scout image (▶**Fig. 60.3**) of the brain shows a linear skull fracture. Bone debris and fragments are observed as well as the skull opening created by the bullet.
   - Things to look for in the CT scan images:
     - Was there any involvement of more than one lobe?
       - Involvement of more than one, lobe especially bihemispheric involvement is related with a poor to dismal clinical outcome.[3]
     - Did the built cross the midline or was there any involvement of the sagittal, transverse, or sigmoid sinuses?
       - Involvement of more than one lobe or any of the venous sinuses means that the injury has been of a major magnitude and that favorable recovery or survival is less likely.
     - Was there any exit wound or is the bullet still inside?
       - The bullet can be a source of infection or toxicity (lead or copper)
       - Although this factor is important in planning treatment, neither entry nor exit wounds were predictors of outcome.[3]
     - Are there any signs of edema or impending herniation?
     - Is the differentiation between the gray and white matter preserved?
     - Is there any expanding hematoma or SAH?
       - Effaced sulci, perilesional edema, expanding hematoma, or SAH on CT scans are all indicators of the necessity for rapid intervention and correction of an underlying process in salvageable cases.
       - Global loss of white–gray matter differentiation is usually a moribund sign.
     - Is there any accompanying cervical spine injury?
       - Cervical spine injury is relatively rare.
       - Routinely ordering cervical spine imaging studies, such as CT scan, for patients who sustained gunshot wound injuries to the head are not warranted in asymptomatic patients.
       - Ordering such investigations is reserved for patients with suspected injuries to the cervical spine who are symptomatic or unconscious.[13]
     - Was there any injury to the base of skull or nasal sinuses that increases the chances of cerebrospinal fluid (CSF) otorrhea, rhinorrhea, infection, or abscess formation[14]?
     - Was there any involvement to eloquent areas, for example, motor, sensory, and visual areas?
       - This finding typically means major loss of function and disability.
     - Is there any active brain tissue coming out of the wound, nose, or ears?
       - This finding typically signifies a continuously evolving increased ICP.
   - All of these questions can give an estimation of the patient's expected deficits and disabilities, should he survive the acute stage.
6. *Should you give prophylactic antibiotics? What are the factors that may raise the incidence of infection in such cases?*
   - Intracranial infection can occur in 25% of patients following civilian gunshot injuries. Most of these occur during the first 30 days of follow-up. Associated major comorbidities can be expected especially in patients with compromised immunity, such as diabetes mellitus.

## Answers (*continued*)

- The most common causative organism is *Staphylococcus aureus*, although other organisms have been found on cultures such as *Acinetobacter* spp.[15]
- Evidence show that early surgical intervention reduces the chance of infection and abscess formation.[16]
- There was no association between the administration of prophylactic antibiotics and the rate of development of CNS infection.[14] Despite this statement, the incidence of CNS infection after gunshot wound injuries to the brain remains elevated and the guidelines on "Management and Prognosis of Penetrating Brain Injury" recommend prophylactic antibiotics for all patients with missile head wounds.[15]
- Antibiotics are recommended especially in the following cases where the incidence of intracranial infection is higher:
  - Presence of osseous or metallic intraparenchymal fragments
  - Projectile trajectory through natural orifices potentially contaminated with bacteria
  - Prolonged hospital stay
- The choice of antibiotics can be influenced by the following factors:
  - Bullet type (civilian vs. military) size and caliber
  - Speed of the bullet or shrapnel
    - Slow speed of bullet (civilian weapon) and presence of shrapnel create dirty wounds and may increase the risk of infection.
    - Fast military bullets are usually hot and are more likely to be sterile.
  - Surface area of contact with tissue: for example, knifes have larger surface areas and hence larger bacterial load [Class III].
- Taking surgical swabs and sending fragments and debris for culture and sensitivity as well as obtaining the input of colleagues from the infectious diseases team will aid in the treatment plan and is of great value.
- See ▶**Table 60.2** for a distribution of the types of infections seen in gunshot injuries to the head.

7. *Describe the mechanism of damage in gunshot injuries to the brain.*
   - There are multiple mechanisms involved in the brain damage in patients with gunshot injury to the head. These are summarized below (see also ▶**Fig. 60.4**, ▶**Fig. 60.5**, ▶**Fig. 60.6**, and ▶**Fig. 60.7**).
   - Primary injury:
     - Caused by the direct penetrating and shearing force of the bullet itself
     - This includes the penetrating bone fragments and debris, and the formation of multiple small channels within the brain parenchyma by bullet fragments.
     - Moreover, the negative pressure generated by the bullet cavity can draw contaminated material into the cavity.[15]
     - The supersonic shock wave injury accompanying the bullet subjects the brain to a high amount of kinetic energy. This wave usually lasts for few microseconds and is thought to cause injury not only along the direct path of the bullet, but also to locations far away from the bullet's path.[1]
     - If these locations include the respiratory centers in the brainstem, this causes apnea and death.
     - Other factors that can contribute to the mechanism of injury include coup and countercoup effects.
   - Secondary injury:
     - Includes brain edema created from the initial impact and its sequelae
     - Late mechanisms of injury include infections, abscess, bullet or fragment migration, late seizures, lead toxicity, and aneurysm formation (the latter is highly suspected if the bullet travels along the path of large intracranial vessels, hence the need for an angiogram in salvageable cases).
   - In cases where the gun is held very close to the head, pressurized gases and debris may cause small secondary channels within the brain parenchyma.
   - Bullets with low energy may flip, spin, or rotate inside the brain and hence may cause a wider path and greater damage. Also, bullets crossing a path involving more than one lobe have grater damaging effects.
   - Accompanying bone fragments and shrapnel debris will also increase the amount of injury sustained by travelling into different parts of the brain parenchyma.
   - The toxic effects of copper, other heavy metals, and chemicals cause harmful and inflammatory reactions within the brain substance.
   - Type of gun and bullets used affect the injury, for example, military guns and missiles are usually larger, faster, and cause more damage and are usually more fatal compared to civilian guns. However, in the previously mentioned series military gunshot wound injuries carried better outcomes in terms of mortality and morbidity probably due to a faster medical response within the battlefields.
8. *What are the indications for surgery and why?*
   - There are many indications for surgery. These include:
     - Controlling the bleeding
     - Decreasing and monitoring the ICP
     - Debridement and removal of bone fragments and debris
     - Cosmesis
   - The majority of surgical procedures are directed toward preventing secondary brain injury.

## Answers (*continued*)

- Surgical timing and type of intervention must be decided based on the individual patient presentation.
- The timing of surgical intervention depends on the patients' hemodynamic stability and presenting neurological status (patients without brainstem reflexes with widely dilated unresponsive pupils and patients who had at least a single episode of cardiac arrest should not undergo surgery).
- Intracranial monitoring can be useful in unconscious patients in whom there is no focus for decompression or no signs of bleeding to evacuate. These monitoring devices can also be therapeutic in the management of elevated ICP.[8]
- In cases of decompressive craniotomy for severe brain edema, a large craniotomy is advised.

9. *What type of intervention would you offer this patient and why? How would you plan it?*
   - One should plan a bifrontal craniotomy in cases where both frontal lobes are involved and unilateral decompression in cases where one hemisphere is involved).
   - Copious amount of irrigation with antibiotics are used by some centers aiming to decrease the chances of infection [class III] and vigorous debridement, removing any devitalized brain tissue and bone fragments.
   - Copper in the bullet might be toxic and the bullet may migrate.[18]
   - Going after deep-seated shrapnel or debris should not be attempted initially as this may cause more harm than good.
   - Surgical intervention may be staged dealing with life-threatening risks initially then staging other treatments such as coiling of traumatic unruptured aneurysms or removing deep-seated bone fragments using stereotactic techniques or image guidance for safer and better outcomes.[19]
   - There have also been reports of head shrapnel injuries treated in a semiconservative way with only primary debridement with favorable clinical outcomes.
10. *What is the expected outcome of such patients?*
    - Patients have been broadly categorized into four major groups based on their presenting GCS which also helped in predicting clinical outcome.
    - First group: GCS = 3
      - Patients in this group with corresponding CT brain images are usually unsalvageable, unless they respond to initial ICP reducing measures. Even if the ICP decreases, they usually have a very poor clinical outcome with survival rates of 0 to 15%.
    - Second group: GCS between 4 and 8
      - Patients in this clinical group also have a poor clinical outcome but mortality rates are better than in the first group. Survival rates in this group were 30%.
    - Third group: GCS between 9 and 12
      - This group has far better survival as well as functional outcome compared to the previous two groups, and should be treated promptly and aggressively in order to decrease the amount of damage as much as possible. Survival rate in this group was around 61%.[20]
    - Fourth group: GCS between 13 and 15
      - These patients usually only suffer from functional sequelae and mortality rates are usually low. Survival rate was 97% among patients with a GCS scores ranging from 13 to 15.[20]
    - Although there are several grading tools that correlate the presenting clinical status of the patient and outcome. The Glasgow Outcome Score (GOS) remains the most widely used and accepted system among them (see ▶**Table 60.3**).
    - The correlation between GOS and basal cisterns on CT brain in traumatic brain injury can be used in these cases as a predictive tool of outcome (see ▶**Table 60.4**).
    - The long-term functional and cognitive as well as neuropsychosocial deficits of gunshot wound injury can be improved by neuropsychological rehabilitation.[21]
11. *What factors related to weapons and ammunition might affect the severity of the injury?*
    - Speed:
      - Low muzzle speed < 250 m/s (most hand guns are in this speed category): injury is mostly caused by primary effects of laceration and penetration.
      - High muzzle velocities of 600 to 750 m/s (military artillery) cause shock wave damage and explosive intracranial injury in addition to primary effects of penetration.
    - Distance: this usually has very small effect on bullet injuries due to the fact that bullets lose a very small portion of their energy crossing long distance.
    - Type of gun and ammunition used: military ammunition are of greater diameter hence causing larger cavities and greater damage.
12. *In penetrating injury to the head due to a stab, when do you attempt to remove the offending object?*
    - In general, one should never attempt to remove a knife or rod causing penetrating injury in the field as it may be tamponading a bleeding vessel.
    - These should be removed in the operating room in a sterile and controlled environment.
    - Stab wound injuries to the brain are usually dirty with higher rates of infection. This is most probably due to the larger surface area of these weapons that comes in contact with tissue (bacterial load), as well as the longer timing they may stay in touch with tissue as compared to bullets which usually have an exit wound.

**Table 60.1** The distribution of gunshot wound injury and shrapnel injury according to location[7]

| | |
|---|---|
| Frontal | 33.75% |
| Temporal | 17% |
| Parietal | 18% |
| Occipital | 9.25% |
| Posterior fossa | 0.25% |
| Orbitocranial | 8.75% |
| Multiple sites | 8.75% |

**Table 60.2** Type and percentage of complications following gunshot wound injury to the brain[14]

| Type of Complication | Percentage |
|---|---|
| Meningitis | 50 |
| Osteomyelitis | 5 |
| Brain abscess | 25 |
| Subdural empyema | 7 |
| Cerebritis | 7 |
| Ventriculitis | 5 |

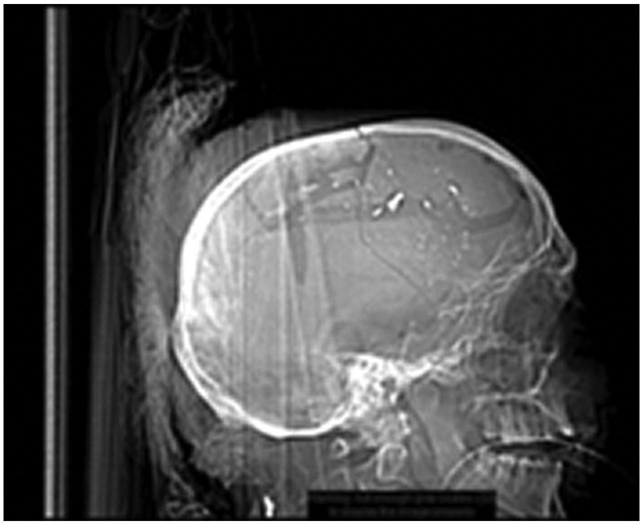

**Fig. 60.3** Sagittal scout image scanogram of patient described herein.

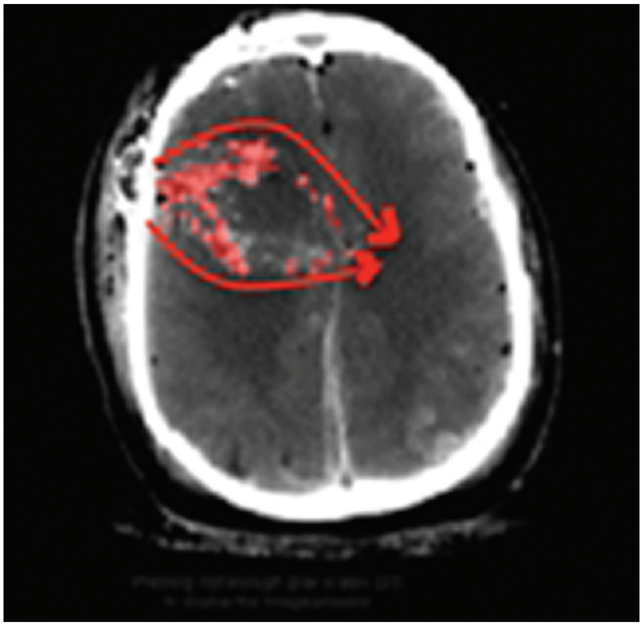

**Fig. 60.4** Axial CT scan without contrast: segmentation of the bullet's path highlighted in *red* on brain CT shows the initial disparity of bone fragments and debris in a transverse "V"-shaped pattern around the bullet cavity created.[17]

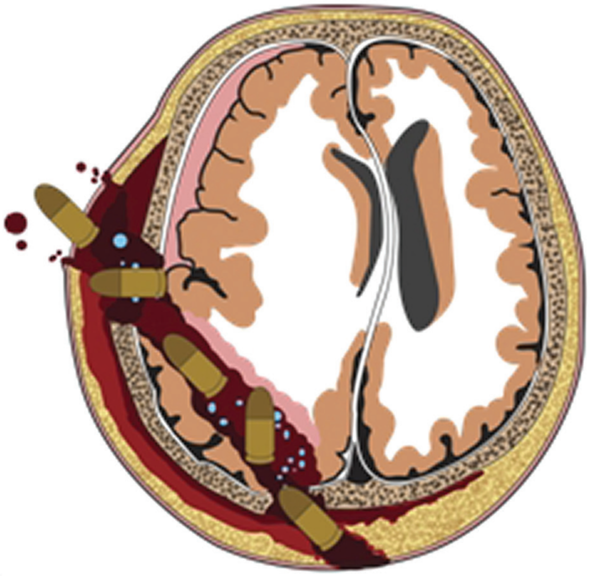

**Fig. 60.5** Artist's rendering illustrating the deviation of the bullet from its path and the rolling-over effect of the bullet that could increase the penetration diameter.

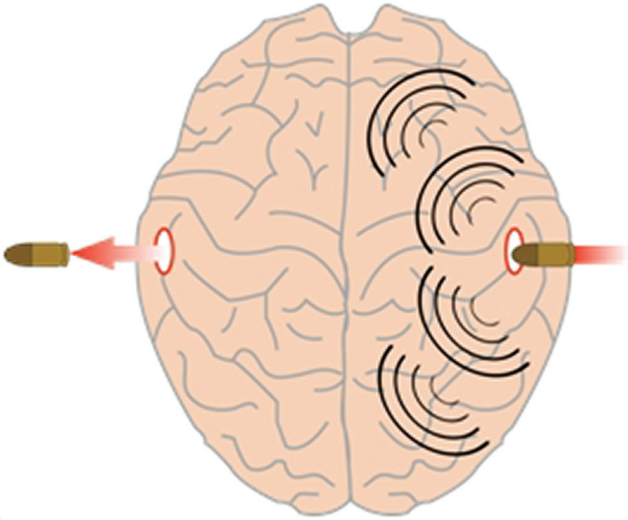

**Fig. 60.6** Artist's rendering illustrating the supersonic shock wave accompanying the bullet injury spreading across different parts of the brain.

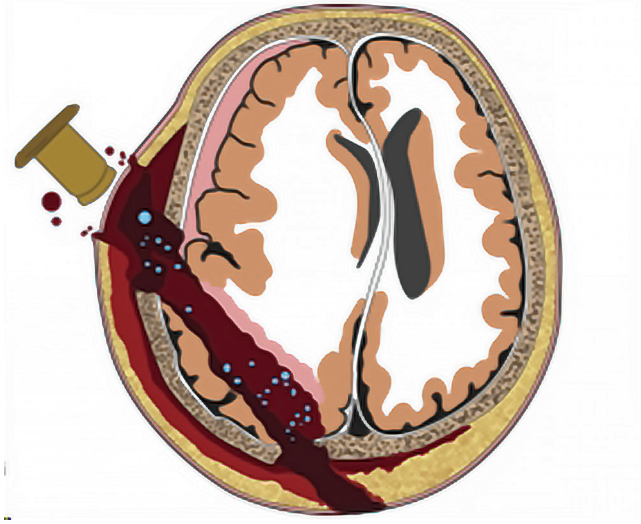

**Fig. 60.7** Artist's rendering demonstrating mushrooming of the bullet upon impact that will increase the penetration diameter and hence the damaging effects.

**Table 60.3** Clinical status of the patient and outcome as per the GOS

| Score | Meaning |
|---|---|
| 5 | Good recovery: resumption of normal life despite minor deficits |
| 4 | Moderate disability (disabled but independent): travel by public transportation, can work in sheltered settings |
| 3 | Severe disability (conscious but disabled): dependent for daily support (may be institutionalized) |
| 2 | Persistent vegetative state: unresponsive and speechless; after 2–3 wk may open eyes and have sleep/wake cycles |
| 1 | Death: most deaths ascribable to primary head injury occurs within 48 hr |

*Abbreviation:* GOS, Glasgow Outcome Score.

**Table 60.4** Correlation between GOS and basal cistern status on CT in predicting outcome in traumatic brain injury[22–25]

| Outcome | | | | | |
|---|---|---|---|---|---|
| Basal Cisterns | Mortality (%) | Vegetative State | Severe Disability (%) | Moderated Disability (%) | Good (%) |
| | (GOS 1) | (GOS 2) | (GOS 3) | (GOS 4) | (GOS 5) |
| Normal | 22 | 6 | 16 | 21 | 35 |
| Compressed | 39 | 7 | 18 | 17 | 19 |
| Absent | 77 | 2 | 6 | 4 | 11 |
| Non visualized | 68 | 0 | 11 | 9 | 125 |

*Abbreviation:* GOS, Glasgow Outcome Score.

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# Case 61 Other Penetrating Intracranial Trauma

*Domenic P. Esposito*

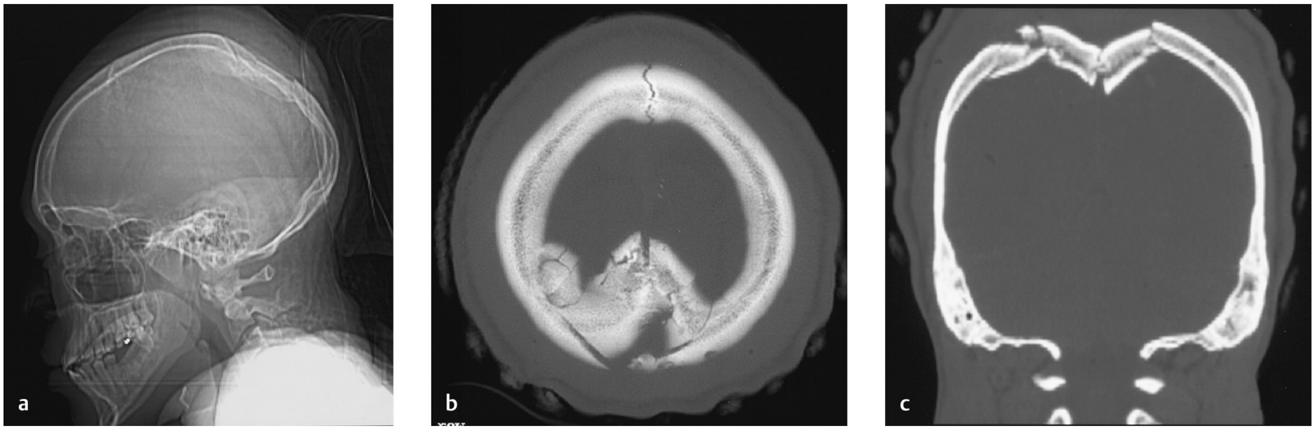

**Fig. 61.1** CT scan of the head with (**a**) scout image, (**b**) axial, and (**c**) sagittal views, showing depressed skull fracture in the occipital area.

## Clinical Presentation

- A 27-year-old man presents to the emergency room (ER) with a history of being struck in the head with a blunt object.
- The initial Glasgow Coma Scale (GCS) score in the ER is 15.
- There are no focal neurologic deficits.
- A CT scan is obtained and shown in ▶**Fig. 61.1.**

## Questions

1. What would be your initial work-up and management plan?
2. Other than radiologic studies, what other studies would be indicated?
3. Outline your plan of care at this point.
4. Interpret the MRI scan shown in ▶**Fig. 61.2**.
5. Following imaging and laboratory studies, the patient develops a right hemiparesis and a speech disturbance. What is the likely cause of these findings?
6. What would be your management plan at this time?
7. What position would you use for the operative approach?
8. What special arrangements would you make preoperatively?
9. Assuming the blunt force trauma produced a very large, somewhat stellate laceration, how would you plan your skin flap?
10. What nontypical neurosurgical devices might be of value in this case?
11. Would you give your anesthesiologist any particular instructions?

    You look at the anesthesiology monitor before beginning (which is usually a good idea) and see that the patient has a pulse of 120 beats/minute, a systolic blood pressure of 90 mm Hg, and the central venous pressure (CVP) line is not connected to a monitor.
12. What is your next step in the management of this patient?
13. The patient is now stable and you open the skin flap revealing the underlying depressed skull fracture, there is no excessive bleeding at this point. Would you begin removing depressed fragments, or would you plan a bone flap (if so, describe the flap)?
14. Before beginning the bone work would you consider harvesting any tissue? If so, what would be some of your choices?
15. While performing your bony removal, copious bleeding begins from a disrupted sagittal sinus. What is your next step in management?

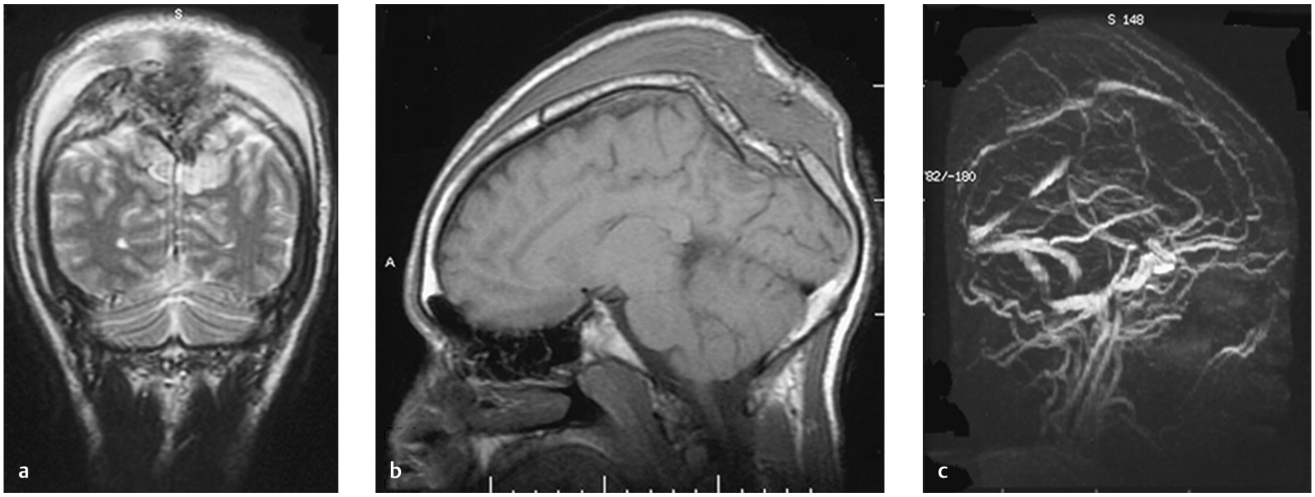

**Fig. 61.2** MRI of the brain with coronal T2-weighted (**a**) and sagittal T1-weighted (**b**) images showing the depressed skull fracture with underlying brain contusions. MR venography (**c**) showing disruption of the posterior portion of the superior sagittal sinus.

## Answers

1. *What would be your initial work-up and management plan?*
   - As the patient is currently stable, laboratory studies should be obtained including a complete blood count, electrolytes, type and screen, and coagulation profile.
   - The patient should be kept in a monitored bed while further treatment is planned.
   - Prophylactic anticonvulsants may be given.
2. *Other than radiologic studies, what other studies would be indicated?*
   - An urgent (STAT) MRI with MR venography (MRV) should be obtained; if MRV is not available a CT venogram (CTV) or angiography with late venous phases would be acceptable.
3. *Outline your plan of care at this point.*
   - Because it is highly unlikely that this lesion can be managed conservatively, preparations for surgical treatment should be initiated. See **Case 47** for details of surgical considerations.
4. *Interpret the MRI scan shown in* ▸**Fig. 61.2.**
   - The MRI reveals a large subgaleal hematoma with compromise of the superior sagittal sinus and cortical contusions.
5. *Following imaging and laboratory studies, the patient develops a right hemiparesis and a speech disturbance. What is the likely cause of these findings?*
   - The patient's neurologic deterioration is most likely due to compromise of the sagittal sinus.
6. *What would be your management plan at this time?*
   - This patient needs to be taken to the operating room for elevation of the depressed fracture and possible repair of the sagittal sinus.[1]
7. *What position would you use for the operative approach?*
   - The patient should be positioned prone with the head slightly elevated.
   - Mayfield three-point rigid fixation may be used.
8. *What special arrangements would you make preoperatively?*
   - Preoperative preparation should include:
     - Type and cross-match at least two units of packed red blood cells
     - Precordial Doppler probe
     - Central line and arterial line
     - Preoperative antibiotics and anticonvulsants
   - This case can very easily turn into a surgical disaster. You should arrange to have competent experienced surgical hands other than your own to assist you.
9. *Assuming the blunt force trauma produced a very large, somewhat stellate laceration, how would you plan your skin flap?*
   - The scalp flap was partially made by the assailant, but extension of the stellate laceration in both anteroposterior and lateral directions for good exposure is mandatory.
10. *What nontypical neurosurgical devices might be of value in this case?*
    - Some nontypical neurosurgical devices that might be of help are fogarty catheters, cell savers, and vascular grafts.
11. *Would you give your anesthesiologist any particular instructions?*
    - The anesthesiologist needs to be aware of the possibility of extensive rapid blood loss and the blood needs to be ready in the room prior to bone removal.
12. *What is your next step in the management of this patient?*
    - After gently reminding anesthesia of the tenuous state of the patient, insist that the patient be adequately transfused, monitored, and stabilized prior to proceeding with the procedure.

## Answers (*continued*)

- Ensure CVP is monitored.
- Ensure the patient is being treated for shock: fluids, blood transfusion, and pressors if necessary.

13. *The patient is now stable and you open the skin flap revealing the underlying depressed skull fracture, there is no excessive bleeding at this point. Would you begin removing depressed fragments, or would you plan a bone flap (if so, describe the flap)?*
    - Removing the depressed fragments without prior exposure and control of the proximal and distal normal sagittal sinus could easily result in the demise of this patient.
    - A circular bone flap around the depressant area with multiple burr holes on either side of the sinus as well as lateral to the fragments is mandatory.[2,3]
14. *Before beginning the bone work would you consider harvesting any tissue? If so, what would be some of your choices?*
    - Harvesting a piece of temporalis fascia to assist with possible reconstruction may be helpful.
15. *While performing your bony removal, copious bleeding begins from a disrupted sagittal sinus. What is your next step in management?*
    - Quickly remove all bone over the sinus. If proper preparations have been made, this can be hastily performed.[3,4]
    - Establish control of the sinus with digital pressure and assess the damage.
    - In this case, a large thrombus was encountered in the distal sinus (which was probably the cause of the patient's neurologic decline) and a 5 cm lateral laceration of the sinus was repaired primarily. The sinus remained intact. This patient eventually made a complete recovery.[5]

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# Section IV Intracranial Pathology: Pediatric Disorders

# Case 62 Aqueductal Stenosis

*Jeffrey Atkinson*

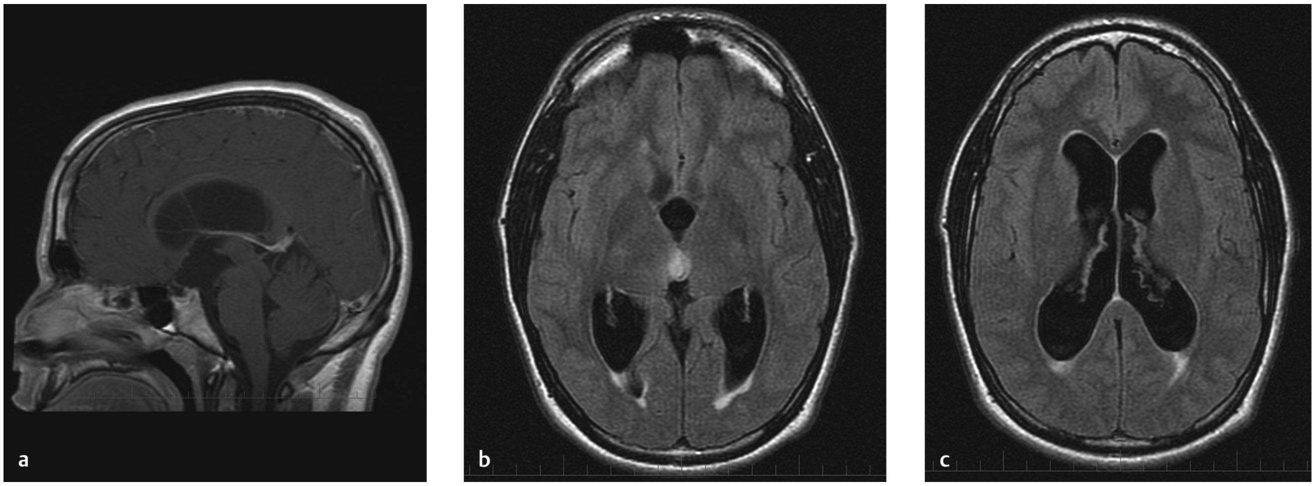

**Fig. 62.1** (**a**) Midsagittal T1-weighted MRI and (**b**, **c**) axial fluid-attenuated inversion recovery image of the brain.

## Clinical Presentation

- A 15-year-old adolescent boy presents with a long history of intermittent syncopal episodes.
- He was initially investigated at age 10 by a neurologist with normal cranial imaging, normal electroencephalogram (EEG), and normal physical examination.
- Similar episodes recurred at age 15 and although his neurologic examination remained normal, MRI was performed as part of further investigation and was significantly different from the prior CT scan.
- Representative images are presented in ▶**Fig. 62.1.**

## Questions

1. Describe the findings on the imaging study.
2. What advice would you give the patient in terms of management?
3. What would be the basis for intervention in this patient?
4. Describe possible surgical interventions for the management of this patient.

   The patient underwent a successful endoscopic third ventriculostomy (ETV) and biopsy of the third ventricular mass. The pathology was consistent with juvenile pilocytic astrocytoma.
5. Describe the risks associated with the above procedure.
6. Are there any specific technical considerations for the procedure?
7. What is the long-term prognosis for this patient?
8. What is your management strategy for the patient given the above information?

## Answers

1. *Describe the findings on the imaging study.*
   - The MRI demonstrates ventriculomegaly at the level of the lateral ventricles with some transependymal edema.
   - There is also an apparent mass in the posterior part of the third ventricle and tegmentum of the brainstem.
2. *What advice would you give the patient in terms of management?*
   - Management strategies for this patient can be divided into observational and interventional strategies.
   - Given the relatively minor history and lack of clear relationship between the lesion and symptoms, some may argue that the patient could be followed closely.
   - However, the change in lesion and ventricles from the scan 5 years previously and the transependymal cerebrospinal fluid (CSF) on MRI, all argue for treatment of hydrocephalus.
3. *What would be the basis for intervention in this patient?*
   - The basis for intervention is the transependymal migration of CSF seen on MRI, and the clear progressive dilation of the ventricles over time, both of which suggest increasing hydrocephalus, and the probable growth of the mass over the time interval between the two imaging studies.
4. *Describe possible surgical interventions for the management of this patient.*
   - This patient could receive CSF diversion by means of a ventriculoperitoneal (VP) shunt.
   - CSF diversion could also be performed by ETV.[1]
   - Consideration could be made for biopsy of the lesion at the time of ETV given the apparent location of the lesion in the posterior third ventricle.[2] It is not very likely that this lesion will progress and thus consideration for resection of this lesion would not be necessary due to absent progressive growth or unfavorable histology on biopsy.
5. *Describe the risks associated with the above procedure.*
   - ETV presents risks of late or early failure, fornix injury, intracerebral hematoma, hypothalamic injury, basilar aneurysm or arterial injury, and uncontrolled bleeding.[3,4]
6. *Are there any specific technical considerations for the procedure?*
   - Technical considerations revolve specifically around the preferred option of ETV with lesional biopsy.
   - It would be essential that the ETV be performed first to make sure that this procedure was not aborted due to bleeding from the biopsy.[2]
   - To perform both the ETV and the biopsy a flexible endoscope would be needed.
   - Alternately, using a rigid scope, consideration must be given to placement of a single, slightly more anterior burr hole or two burr holes to allow an appropriate trajectory to both the anterior third ventricle and ventriculostomy site and the posterior third ventricle for the biopsy.
7. *What is the long-term prognosis for this patient?*
   - Expectations for the long-term control of hydrocephalus in this patient are good, approximately 80%.
   - Typically, tegmental, posterior third ventricle lesions of this type are benign and slow growing, and cause no further problems apart from the hydrocephalus.
   - If a biopsy was performed, this may further define prognosis.
8. *What is your management strategy for the patient given the above information?*
   - ETV with or without biopsy would be the best option followed by serial imaging observation of the mass lesion.[2,5]

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# Case 63 Cerebrospinal Fluid Shunt Infections

*Jeffrey Atkinson*

## Clinical Presentation

- A 3-month-old child with an L5-level myelomeningocele and a ventriculoperitoneal (VP) shunt presents to the emergency department with 24 hours of progressive irritability and fever.
- The child had the spinal defect closed at 2 days of life and a VP shunt inserted for progressive macrocrania and hydrocephalus at 14 days of life.
- He has been on a program of home intermittent catheterizations since birth and no prophylactic antibiotics.
- The exam demonstrates child with a fever of 39.5°C, bulging fontanel, somnolent, and irritable.
- The motor examination is unchanged with no plantar flexion in the feet, but otherwise normal.
- The child's incisions all look well healed.
- White blood cell (WBC) count is elevated, and the urinalysis shows positive bacteria and WBCs.

## Questions

1. What is the differential diagnosis suggested by this child's presentation?
2. What investigations are appropriate and why?
   Blood cultures are collected and urine cultures are sent. A CT scan is done which shows a stable ventricle size compared with the last scan done after the shunt was inserted. Shunt tap reveals 1500 WBCs and 10 red blood cells (RBCs) with no bacteria seen on a gram stain.
3. What is the diagnosis?
4. What are the usual organisms involved?
5. What are the treatment options for this child?
6. What antibiotic regimen would you choose?
7. What is the incidence of shunt infection after an initial procedure?
8. What is the time frame over which these infections usually develop?
9. What is the incidence of shunt infection after an initial shunt infection?
10. What maneuvers have shown benefit in reducing shunt infection?

## Answers

1. *What is the differential diagnosis suggested by this child's presentation?*
   - This child is presenting with the clinical signs of an infectious illness, and clinical signs of shunt malfunction, though the fever itself may be causing the child's irritability independent of shunt malfunction, and a severely irritable child may present with a bulging fontanel.
   - The cause of the febrile illness may be viral or respiratory, but in this case, we would be worried about shunt infection or wound complication, as well as urosepsis.
2. *What investigations are appropriate and why?*
   - This child needs cultures of blood, urine, a complete blood count, chest X-ray as well as viral swabs of any upper respiratory tract-related secretions and a full exam to look for skin abrasions, wound complications, or other pertinent physical findings.
   - CT scan is indicated due to the clinical signs of increased intracranial pressure (ICP).
   - A tap of the shunt reservoir to obtain cerebrospinal fluid (CSF) is clearly indicated in this child given the presentation and relatively recent shunt surgery.
3. *What is the diagnosis?*
   - The cell count of the CSF points toward shunt infection even in the absence of an initially positive gram stain.
   - The culture of CSF will probably grow eventually.
4. *What are the usual organisms involved?*
   - Shunt infections are usually from skin colonization occurring at the time of surgery—gram-positive cocci are most common.[1]
   - In a child of this age, and especially one with urinary catheterizations, gram-negatives and coliforms are also possible.[2]
5. *What are the treatment options for this child?*
   - This child needs antibiotics and some type of shunt externalization procedure.
   - Very few shunt infections will respond to antibiotics alone without hardware removal.

## Answers (*continued*)

- Many centers will externalize the shunt by removing the distal catheter from the abdomen and then treat until the CSF is sterile before replacing the entire system.[1]
- In the event of continued positive cultures, the entire shunt system should be converted to an external ventricular drain (EVD).[2]
- Some centers would externalize the EVD upfront with removal of the whole shunt system.[1,2]
- The duration of antibiotic treatment before reinternalization is debatable; it averages 10 to 14 days, but three consecutive negative CSF cultures is a common standard.[2,3]

6. *What antibiotic regimen would you choose?*
   - Initial antibiotic regimen needs to include broad-spectrum CSF penetrating coverage, and good antistaphylococcal coverage until the organism is known.[1,2]
   - Typical initial regimens would include a third-generation cephalosporin with vancomycin plus or minus an aminoglycoside.
7. *What is the incidence of shunt infection after an initial procedure?*
   - Shunt infection rates per procedure after an initial shunt placement are approximately 8 to 10% in most large studies of shunt insertions in children.[1]
8. *What is the time frame over which these infections usually develop?*
   - Most shunt infections are procedure-related and present within the first 6 months of surgery.
   - Other risk factors for shunt-related infection include wound breakdown and CSF leak.[1]
9. *What is the incidence of shunt infection after an initial shunt infection?*
   - Shunt infections may occur up to 25% of the time when a shunt is replaced after an initial infection.[1]
10. *What maneuvers have shown benefit in reducing shunt infection?*
    - There are a great many studies attempting to demonstrate protocols to reduce shunt infection rates.[4]
    - Efforts to reduce shunt infection rates have included perioperative antibiotics, various procedure-related technical issues such as double gloving, short duration of surgery, surgery timing, reduced mechanical manipulation of the hardware, and more recently antibiotic-impregnated shunt catheters.[4–7]
    - Many of these techniques, including antibiotic-impregnated catheters, are still the subject of some debate as to their relative efficacy.[4,6,7]

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# Case 64 Slit Ventricle Syndrome

*Iván Verdú-Martínez, Pablo González-López, and Samer K. Elbabaa*

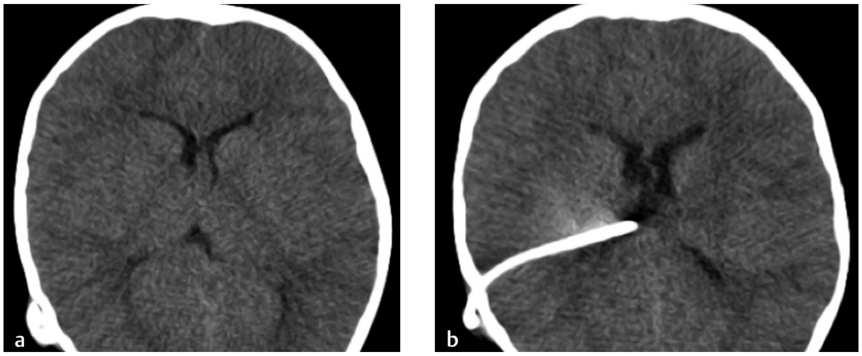

**Fig. 64.1** **(a, b)** Brain CT scan revealing relatively well-decompressed small lateral ventricles and ventricular shunt catheter within the right lateral ventricle.

## Clinical Presentation

- A 2-year-old boy is presenting with headaches and vomiting. A ventriculoperitoneal (VP) shunt was inserted shortly after birth during the perinatal period due to a history of intraventricular hemorrhage 3 days after birth.
- The family mentions a history of three previous shunt revisions during the last year.
- He complains of new onset disabling headaches and vomiting over the past several weeks. Neurological exam is intact.
- CT scan shown in ▶**Fig. 64.1** demonstrates well-decompressed small ventricles compared to previous CT scans.

## Questions

1. Describe the imaging findings.
2. How would you define the slit ventricle syndrome?
3. Do you know any pathophysiological explanation for this syndrome?
4. What should be the first step in the management of this patient?
5. What are the next steps to be followed should the initial steps be inconclusive?
6. What are the surgical treatment options?
7. In which patients is lumboperitoneal shunt contraindicated?
8. What should be your treatment options for the patients described in question 7?
9. Is there any classification for patients suffering from headaches and having a shunt?
10. Is there any association with acquired craniosynostosis?

## Answers

1. *Describe the imaging findings.*
   This is an axial view of a CT scan with small decompressed ventricles and a VP shunt placed in the right occipital horn. No other abnormalities can be identified.
2. *How would you define the slit ventricle syndrome?*
   - An accepted definition for this syndrome is the combination of intermittent headaches in a shunted patient and small ventricles on imaging by CT or MRI scans.
   - In some articles, there is a measure used to classify the ventricular size known as the fronto-occipital horn ratio (FOHR) which consists of the division of the average of the maximum width of the frontal and occipital horns, by the lateral diameter of the skull.
     - The normal value is considered more than 0.37.
     - Kan et al proposed a FOHR < 0.2 as a reference value to define slit ventricles.
3. *Do you know any pathophysiological explanation for this syndrome?*
   - The most accepted explanation for such symptoms is an overdrainage that provokes ventricular collapse and, subsequently, proximal catheter obstruction and malfunction. This theory could explain the intermittent behavior of the headaches, because the shunt starts to work after a brief period of malfunctioning, due to the enlargement of the ventricles.
   - Rekate et al described obstruction at the foramen of Monro after shunting due to CSF flow inversion. This theory would be useful to explain high intracranial pressure (ICP) symptoms in patients with normal or slit ventricles.
   - Classically, it has been thought that the absence of ventricular enlargement may be due to a diminished brain compliance in those patients. However, Shapiro et al showed that the pressure-volume index is normal in patients who suffer from symptoms of slit ventricle syndrome. Engel et al concluded that the periventricular gliosis present in these patients could lead to an expansion failure of the ventricles with the subsequent increasing of the ICP, followed by a worsened neurological status. However, the fact that some of these patients treated with a lumboperitoneal shunt showed ventricular enlargement after its insertion, goes against this theory.
   - Benzel et al proposed that the increase of the venous transmural pressure in these patients could be the cause for an increased venous congestion and higher brain elasticity that finally leads to an inability of the ventricles to expand.
   - Another study by Rekate et al proposed that the increase of brain turgor due to an obstruction of the venous outflow could be the explanation for the difficulty of the ventricular system to reach a normal size.
   - Albright et al suggested that the cause of ICP increase is related to the premature closure of skull sutures because of lack of skull growth stimulus. In shunted patients, a decrease of the CSF volume, normally serving as skull growth stimulus pulsations, is found. This theory could explain why some symptoms of slit ventricle syndrome patients can improve after a decompressive craniectomy. A recent hypothesis to explain the dissociation between low intraventricular pressure and high ICP is based on capillary "laziness."
4. *What should be the first step in the management of this patient?*
   - The first step is to study the shunt system via imaging (CT/MRI and shunt series X-rays) and consideration of shunt tap to rule out shunt malfunction. At this point, following two possibilities exist:
     - A working shunt with likely overdrainage (can be managed by increasing the shunt resistance via upgrading the shunt valve into a higher-pressure valve in case of fixed pressure valve, or via upgrading the pressure into a higher value in case of programmable shunt valve)
     - A nonfunctioning shunt (should be managed by shunt exploration and revision)
   - In our patient's case, we found a working shunt, so we chose to upgrade the programmable shunt pressure without improvement of symptoms and no changes in ventricular size.
5. *What are the next steps to be followed should the initial steps be inconclusive?*
   - ICP monitoring becomes mandatory in these patients to assess if there is an intracranial hypo- or hypertension.
   - External ventricular drain (EVD) placement can be used as a diagnostic and therapeutic tool simultaneously.
6. *What are the surgical treatment options?*
   - Shunt replacement to a programmable valve with an antisiphon device to prevent overdrainage could be appropriate.
   - An endoscopic third ventriculostomy (ETV) in case where the ventricular anatomy is favorable
   - Lumboperitoneal shunt insertion if there is communicating hydrocephalus, with or without a programmable valve. One should also consider the possibility of performing an ETV first (to convert noncommunicating hydrocephalus into communicating) and then placing a lumboperitoneal shunt.
   - In cases of uncontrolled intracranial hypertension, a decompressive subtemporal craniectomy is the treatment option of last resort.

## Answers (*continued*)

7. *In which patients is lumboperitoneal shunt contraindicated?*
   - Chiari types I and II malformations
   - Tethered spinal cord
   - Those with hydrocephalus related to achondroplasia due to severe spinal stenosis
8. *What should be your treatment options for the patients described in question 7?*
   - The use of a cisterna magna–peritoneal shunt is described in the literature to prevent the complications related to a lumboperitoneal shunt in this group of patients.
9. *Is there any classification for patients suffering from headaches and having a shunt?*
   - Rekate et al proposed five different mechanisms to explain the symptoms present in this syndrome:
     - Low ICP headache
     - Intermittent proximal catheter obstruction
     - Shunt malfunction with small ventricles
     - Intracranial hypertension with working shunt
     - Nonrelated headache
10. *Is there any association with acquired craniosynostosis?*
    - Yes, a premature skull sutural closure can be seen because of the effect of chronic shunting in these patients. The reduction of cranial vault size and acquired limited growth can cause a rise in ICP.

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# Case 65 Mega-hydrocephalus

*Maqsood Ahmad and Abdulrahman J. Sabbagh*

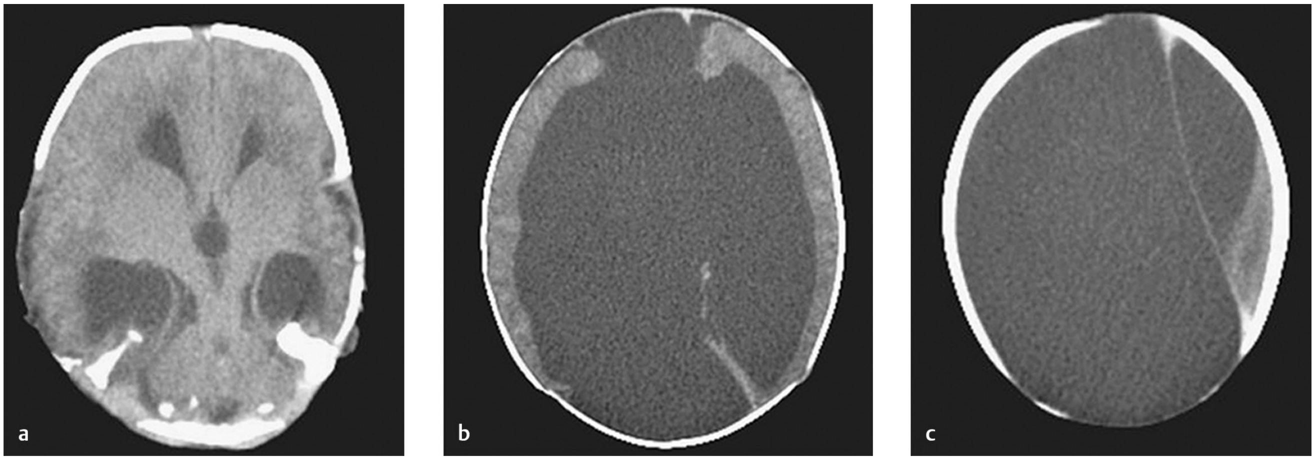

**Fig. 65.1** CT scan, axial section through the fourth ventricle (**a**), and higher cuts (**b**) and (**c**).

## Clinical Presentation

- You are called to see a newborn in the neonatal intensive care unit diagnosed prenatally with severe hydrocephalus.
- The patient was delivered at 38 weeks of gestation through an elective cesarean section with normal appearance, pulse, grimace, activity, respiration (APGAR) scores.
- On exam, he had a head circumference that is 4 cm above the 97th percentile, bulging fontanel, and splayed sutures. In addition, distended scalp veins were present.
- Neonatal reflexes were present and he was spontaneously moving all limbs.
- He had no signs of spinal dysraphism or any skin manifestation of syndromic features.

## Questions

1. Describe the CT findings (▶**Fig. 65.1**).
2. What are the surgical options?
3. What would be your timing of surgery?
   You decide to place a ventriculoperitoneal (VP) shunt.
4. What are the shunt-type options that may be appropriate for this patient? What are the determinants that will help you to choose?
5. What are the complications specific to this patient that you will discuss with the parents?

   You manage to insert a VP shunt and use a medium pressure valve.
   The patient does well and gets discharged home within a few days. You see him in clinic at 2 and 6 weeks postoperatively and he seems to be doing

## Questions

fine. Three months after discharge, he presented to the clinic with a fluctuant subgaleal collection that is increasing in size upon coughing, crying, or simply laying in the supine position. You opt for observation for 1 week, but it continues to grow.

6. What do you think is happening now?
7. What are your surgical options?

You admit this child and decide to revise the shunt, downgrade the pressure setting, and repair the leak site (from the original burr hole) with fascia and tissue sealant. The patient does very well postoperatively. He is discharged home without leakage or fluid collection. Along with his follow-up with the pediatrics service for delayed milestones, he follows up with you for the VP shunt. You notice that he has started to develop progressively significant positional plagiocephaly.

8. What are the management options?

Despite multipositional stimulation, the head continues to deform upward; the parietal bones grow and override the flattened occipital and frontal bones (▶**Fig. 65.2**). This deformity worsens with time. The fontanel remains soft. There are no associated neurologic sequelae.

At the age of 1 year, the parents are offered surgery, which they opt for. Multiple cranial osteotomies and reconstruction (cranial reduction procedure) are performed to reduce the towering of the cranium, expand the anteroposterior and bilateral diameter, and correct the left-sided occipitoparietal flattening.

9. What was the purpose of a craniofacial reduction procedure in this child?
10. What are the limitations of craniofacial reduction procedures?
11. What complication risks are associated with this elective procedure?

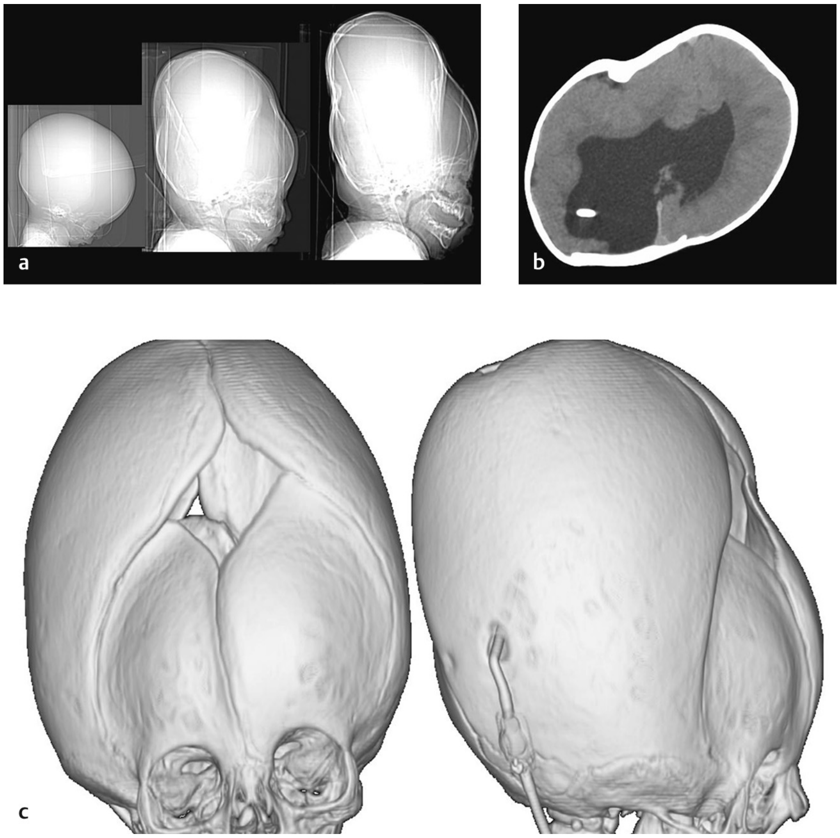

**Fig. 65.2** Postshunting scout lateral images **(a)** axial CT scan through the ventricles **(b)** and three-dimensional reconstructions of CT scan **(c)**.

## Answers

1. *Describe the CT findings* (▶**Fig. 65.1**).
   - This study depicts severe hydrocephalus with significantly thinned out cortical mantle.
   - The fourth ventricle is small, and this represents a case of aqueductal stenosis.
   - There is interhemispheric transcallosal schizencephaly.
2. *What are the surgical options?*
   - Surgical options include:[1,2]
     - Extracranial cerebrospinal fluid (CSF) diversion procedures:
       - VP shunt
       - Ventriculoatrial shunt
     - Third ventriculostomy with or without choroid plexus coagulation[3]
3. *What would be your timing of surgery?*
   - As soon as the patient is medically fit for surgery, i.e., in the first few days of life
4. *What are the shunt-type options that may be appropriate for this patient? What are the determinants that will help you to choose?*
   - Shunt types include programmable versus fixed (low or medium) pressure valves, with or without an antisiphon device.[1,2]
   - The pressure to be used will be determined during surgery from the opening pressure of the ventricle.
5. *What are the complications specific to this patient that you will discuss with the parents?*
   - Specific complications include:[4–6]
     - CSF leakage around the ventricular catheter, as the cortical mantle is very thin in this case. The CSF can track through the burr hole. In addition, Laplace's law describing pressure difference over an interface in a sphere will dictate that the greater pressure in a larger skull will tend to drive further CSF out of the burr hole.
     - Overdrainage from the shunt
     - Shunt infection
     - Pressure sores, valve erosion through the skin, skin abrasions at the overriding bone edges
     - Cranial deformities due to overriding of the floating bones that were much larger than needed preshunting
     - Fluid and electrolyte imbalance due to the CSF shifts during surgery
     - Occult subdural hemorrhage postoperatively: always beware of subdural hematomas developing post shunting, especially in patients with mega-hydrocephalus. These patients will not show signs of high intracranial pressure. They may lose a large portion of their blood volume intracranially and present with hypovolemic shock.
     - General shunt complications and complications of neonatal anesthesia
6. *What do you think is happening now?*
   CSF is leaking from the burr hole site around the ventricular catheter insertion site due to a lower resistance than that at the shunt valve.
7. *What are your surgical options?*
   - Management of a subgaleal collection post shunting:
     - Revise the shunt reservoir to a lower setting and repair the CSF leak site.
     - Repair the CSF site without revising the valve pressure. In this case, there is a risk of recurrence.
8. *What are the management options?*
   - Postshunting plagiocephaly treatment options include:[7,8]
     - Conservative options:
       - Multipositional stimulation
       - Frequent position changes—special care not to lie on the flat areas
       - Correction bands and helmets
   - Special care not to develop abrasions from overriding skull edges
   - Pressure sores at the valve and catheter sites—tailored openings in helmets at these sites
   - Surgical: correction of deformities and remodeling
9. *What was the purpose of a craniofacial reduction procedure in this child?*
   - The purpose of surgery is to improve and ease handling, hygiene, cosmesis, and possibly mobility.[9–11]
10. *What are the limitations of craniofacial reduction procedures?*
    - Limitations include:[9–11]
      - Incapability to reconstruct or change the skull base
      - Presence of a long superior sagittal sinus
      - Risk of infolding of the thinned-out cortex leading to congestion or venous infarct
11. *What complication risks are associated with this elective procedure?*
    - Complications of cranial reduction in craniofacial reconstruction procedures include:[9,10]
      - Hemorrhage and complications of massive transfusion
        - Disseminated intravascular coagulation and other coagulopathies
        - Acute respiratory distress syndrome
      - Postoperative prolonged edema that may threaten the airway and prolong the intubation period
      - Extradural collections
      - Complications of prolonged open surgical procedure
        - Anesthesia issues
        - Pressure sores

## Answers (*continued*)

        - Infection
        - Electrolyte imbalances
    - Risk of brain injury
        - Enfolding of excess cortex causing venous infarcts
        - Manipulation of the superior sagittal sinus and risk of thrombosis or hemorrhage
        - Direct brain injury

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# Case 66 Cerebellar Medulloblastoma

*Philippe Mercier and Frederick Boop*

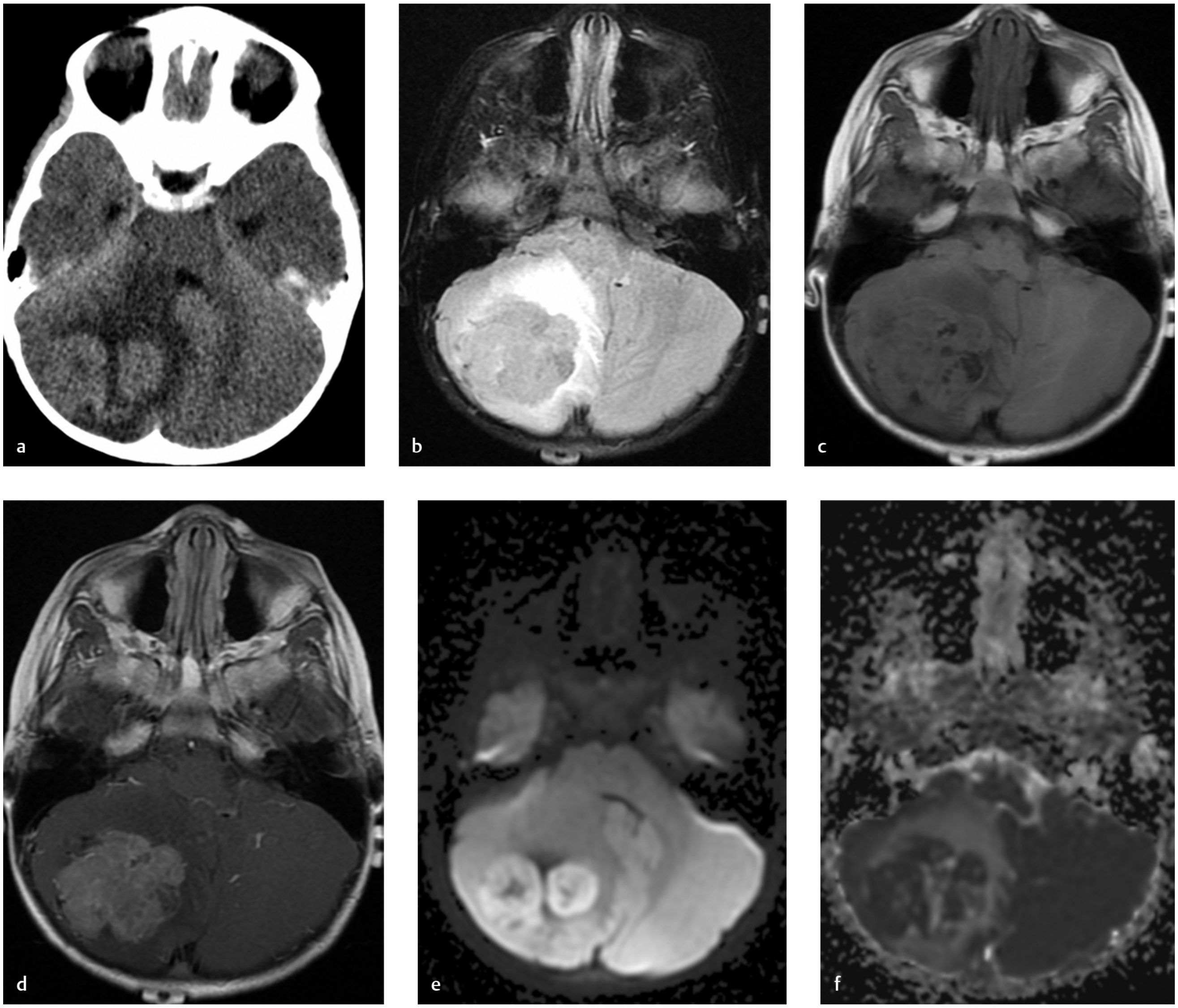

**Fig. 66.1** Axial CT scan of the head demonstrates a mass centered in the right cerebellar hemisphere with mass effect on the fourth ventricle (**a**). The lesion is T2 hyperintense containing multiple internal cysts and a small volume of blood products (**b**). T1 axial MRI without (**c**) and with contrast (**d**) demonstrate solid enhancement. The lesion is diffusion-weighted imaging (DWI) light (**e**) and apparent diffusion coefficient (ADC) dark (**f**) suggesting diffusion restriction.

## Clinical Presentation

- A 3-year-old male presents with 2 months of headache and more recent 2-week history of vomiting and lethargy.
- On exam, the child is awake, alert, and will answer questions appropriately. Eyes are open spontaneously with pupils that are equal and reactive to light and full extraocular movements.
- He has normal strength and appears intact to light touch (▶**Fig. 66.1**).

## Questions

1. What is the differential diagnosis?
2. What other imaging would you recommend?
3. What genetic syndromes are associated with medulloblastoma?
4. How are medulloblastoma classified pathologically?
5. What is the cellular origin of medulloblastoma?
6. Are there any differences in the molecular characteristics of fourth ventricular and parenchymal medulloblastoma in the posterior fossa?
7. What is the role of surgery in the management of medulloblastoma?
8. What is the role of radiation in the treatment of medulloblastoma?
9. Is adjuvant treatment recommended for medulloblastoma?
10. How often is ventriculoperitoneal shunt required in posterior fossa medulloblastoma?

## Answers

1. *What is the differential diagnosis?*
    - Medulloblastoma, pilocytic astrocytoma, ependymoma, atypical teratoid/rhabdoid tumor, ganglioglioma, metastasis (although rare in children)
2. *What other imaging would you recommend?*
    - MRI with and without contrast is required to completely evaluate for drop metastases that typically appear as sugar coating of the dura and nerve roots. Medulloblastoma seeds to the spine in up to half of the cases.[1] The incidence of spinal metastases upon presentation for ependymoma is 5%.[2] In the largest case series to date, 6% of low-grade gliomas had metastatic disease either at diagnosis or in follow-up.[3]
3. *What genetic syndromes are associated with medulloblastoma?*
    - Nevoid basal cell carcinoma syndrome (Gorlin's syndrome)
    - Turcot's syndrome A
    - Li–Fraumeni syndrome
    - Rubinstein–Taybi syndrome
4. *How are medulloblastoma classified pathologically?*
    - In previous iterations of the World Health Organization (WHO) classification of tumors of the nervous system, medulloblastoma was always considered grade IV and classified based on histologic patterns (classic, desmoplastic/nodular, medulloblastoma with extensive nodularity, large cell-anaplastic).
    - A recent revision of the WHO classification of medulloblastoma includes four accepted molecular subtypes including the WNT-activated, sonic hedgehog (SHH)-activated, and the group 3 and group 4 that when combined with histologic subtype have improved prognostic and therapeutic significance.[4]
5. *What is the cellular origin of medulloblastoma?*
    - The origin of medulloblastoma also depends on molecular subtype. WNT-activated tumors develop from WNT expressing cells in the lower rhombic lip.
    - SHH-activated tumors arise from SHH modulated cerebellar cortex.
    - The origin of group 3 and 4 medulloblastoma is unclear.[5]
6. *Are there any differences in the molecular characteristics of fourth ventricular and parenchymal medulloblastoma in the posterior fossa?*
    - WNT-activated tumors typically localize to the cerebellar peduncle/cerebellopontine angle cistern.
    - SHH-activated tumors within the cerebellar hemisphere
    - Group 3 and 4 medulloblastoma are localized predominantly within the fourth ventricle.[6]
7. *What is the role of surgery in the management of medulloblastoma?*
    - Recent treatment algorithms divide patients with medulloblastoma into average- and high-risk patients with high-risk features defined as age < 3, presence of residual disease > 1.5 $cm^2$, and the presence of metastases.
    - Although the goal of surgical resection is accepted to be maximal safe surgical resection, obtaining tissue for histological and molecular studies in addition to opening of CSF pathways are also key goals accomplished during surgery.
    - It is important to be aware that postoperative morbidity can be attributed to aggressive resection of posterior fossa medulloblastoma that may lead to posterior fossa syndrome with temporary or permanent cognitive and speech deficits and ataxia.
    - In addition, fourth ventricular tumors are often adherent to the brainstem and their resection is associated with significant morbidity.[7]
    - Recent studies that take molecular subgroup into analysis questioned the prognostic significance of gross total resection for some subtypes of medulloblastoma.[7] When accounting for molecular subgroups, WNT-associated, SHH-associated, and group 3 show no progression-free or overall survival benefit when comparing near total and gross total resection. There is advantage of gross total resection versus subtotal resection for grade 4 tumors with regards to progression-free survival but not overall survival.[7]

## Answers (*continued*)

8. *What is the role of radiation in the treatment of medulloblastoma?*
   - Craniospinal radiation is the standard of care in medulloblastoma but is typically reserved for children of age 3 and older. Typical treatment consists of photon craniospinal radiation at a reduced dose of 2,340 cGy with a posterior fossa boost of 55.8 Gy for standard-risk patients, with high-risk patients of age 3 or older receiving a higher dose of craniospinal radiation of 3,600 cGy with a posterior fossa boost.
   - With the advent of molecular typing of medulloblastoma, new criteria have been proposed to stratify patients risk based on molecular and histological subtype in addition to the presence of metastasis that will determine the radiation treatment regimen.[4,8]
   - Given the improved prognosis of medulloblastoma with current treatment regimens and the sensitivity of young children to the late effects of photon radiation, age is a consideration with a tendency to withhold radiation in children less than 3. There have also been several studies that suggest proton therapy has a lower risk of late complication when compared to photon therapies, which may result in proton therapy becoming more common in the future.[9,10]
9. *Is adjuvant treatment recommended for medulloblastoma?*
   Adjuvant chemotherapy has an important role in the treatment of medulloblastoma with protocols for young children below the age of 3, in standard- and high-risk patients.
10. *How often is ventriculoperitoneal shunt required in posterior fossa medulloblastoma?*
    - Approximately 70 to 90% of patients with posterior fossa tumors present with hydrocephalus. Placement of an external ventricular drain is often essential to stabilize patients prior to complete evaluation and can be beneficial to control intracranial pressure at the time of surgery.
    - The Canadian Preoperative Prediction Rule for Hydrocephalus identifies age less than 2, preoperative papilledema, moderate to severe hydrocephalus, cerebral metastases, medulloblastoma, ependymoma, or dorsally exophytic glioma as risk factors that increase the chance that patients will have persistent hydrocephalus post resection.[11]

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# Case 67 Brainstem Glioma 1—Pons

*Abdulrahman J. Sabbagh, Ayman Abdullah Albanyan, Mahmoud AlYamany, Reem Bunyan, Ahmed T. Abdelmoity, and Lahbib A. Soualmi*

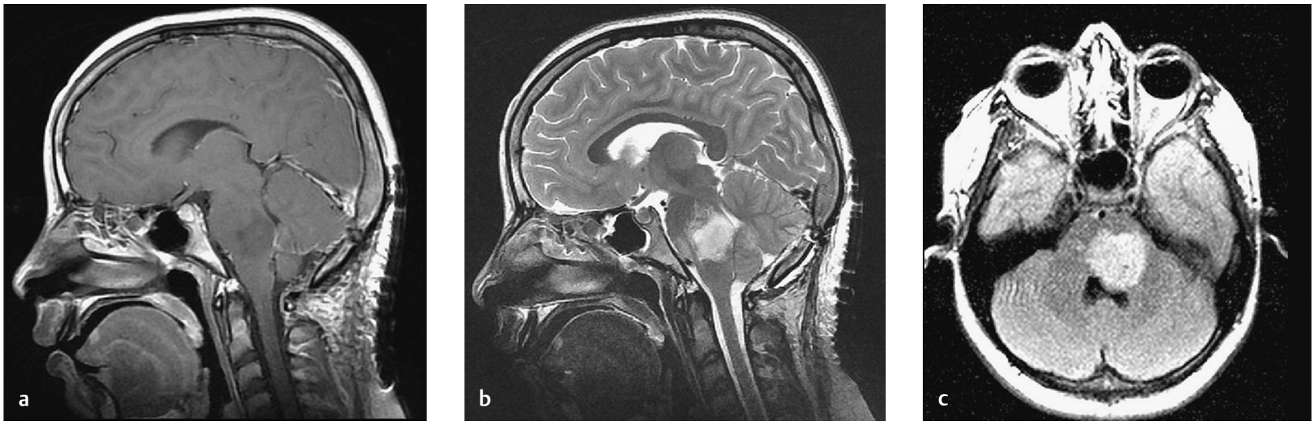

**Fig. 67.1** (**a**) Brain sagittal T1-weighted MRI with gadolinium, (**b**) sagittal T2-weighted MRI and (**c**) axial fluid-attenuated inversion recovery image through the pons.

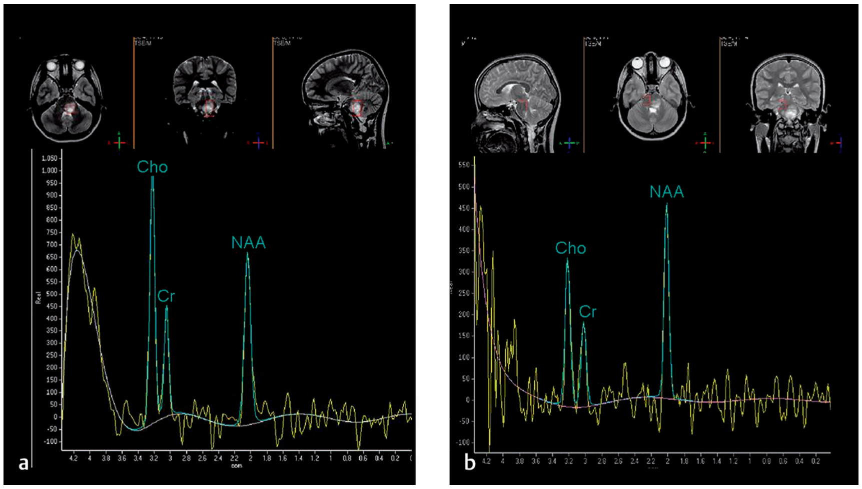

**Fig. 67.2** Magnetic resonance spectroscopy showing voxel configuration taken within the tumor (**a**) and within normal pons (**b**).

## Clinical Presentation

- A 13-year-old right-handed girl presents to a neurologist with slowly progressive double vision and facial asymmetry.
- She complains of some balance problems toward the left side.
- Examination shows a partial left-sided sixth and seventh cranial nerve (CN VI, CN VII) palsy and generalized hyperreflexia. She has normal motor power but a mild right-sided pronator drift.
- An MRI of the brain (▶**Fig. 67.1**) and an MR spectroscopy (▶**Fig. 67.2**) are done. The initial working diagnosis is of a demyelinating process and the patient is started on corresponding treatment.

## Questions

1. Describe the MRI.
2. Describe the MR spectroscopy images. What is your differential diagnosis?
3. How can you anatomically explain the CN VI and VII palsies?

   Over the following months, her diplopia becomes worse and on examination, her CN VI and VII palsies becomes complete and now she has a significant pronator drift and mild swallowing difficulties. A repeat MRI study shows that the lesion is enlarging.
4. If you chose to operate, what would be the aim of the surgery?
5. How would you approach this lesion?
6. What neurophysiologic modalities would you utilize during surgery?
7. What are the safe entry zones into the floor of the fourth ventricle?

   You were able to resect close to 60% of the tumor (▶**Fig. 67.3** shows the postoperative MRI study). The surgery is performed in the intraoperative MRI (iMRI) suite using intraoperative neurophysiology monitoring (IOM) (IOM electrodes were tested for MRI compatibility).

   Her swallowing ability returns to normal, but she has some balance issues that improve with physiotherapy. The CN VI and VII palsies remain. Pathologic tissue diagnosis comes back as grade II diffuse astrocytoma.
8. How would you further manage this case?
9. Classify pontine tumors.

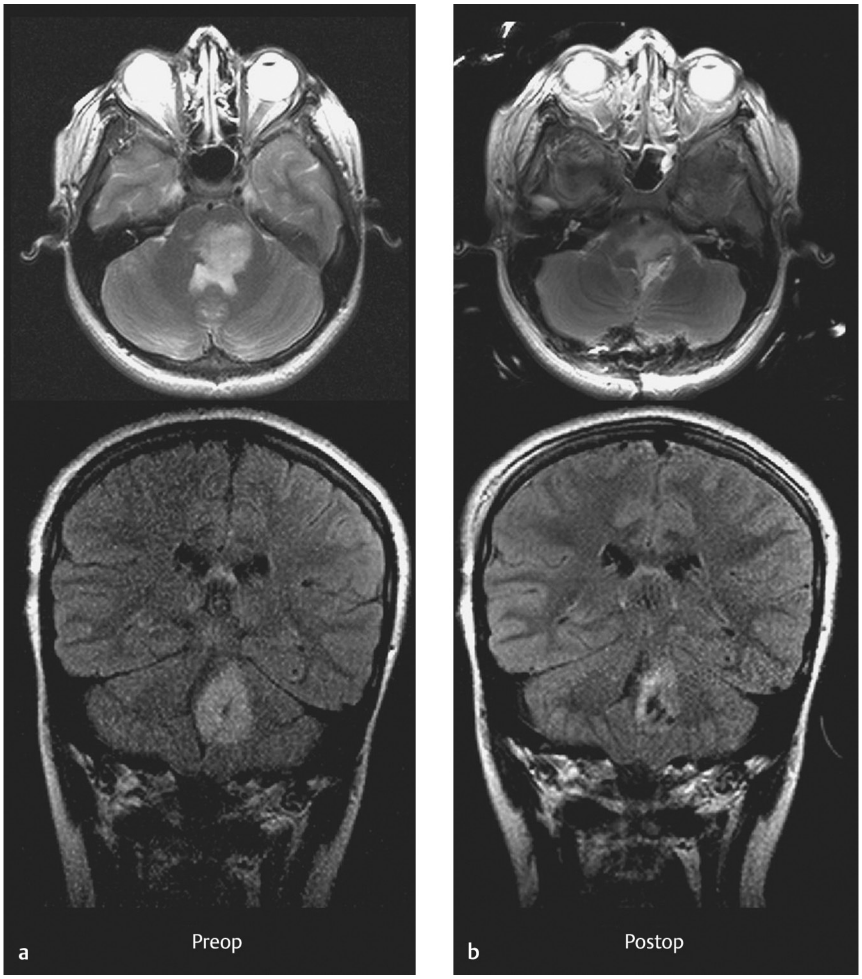

**Fig. 67.3** Comparison between (**a**) preoperative and (**b**) postoperative MRI showing axial T2-weighted and coronal fluid-attenuated inversion recovery images taken at the level of the pontine tumor.

## Answers

1. *Describe the MRI.*
   - There is a nonenhancing hypointense on T1-, hyperintense on T2-weighted images, partly defined pontine lesion that is associated with some edema and deformation of the pons.
   - Fluid-attenuated inversion recovery (FLAIR)-weighted image (▶**Fig. 67.1**c) shows the pontine lesion occupying the right posterior quadrant of the pons, pointing toward the fourth ventricle.
2. *Describe the MR spectroscopy images. What is your differential diagnosis?*
   - MR spectroscopy shows increased choline/creatine (Cho/Cr) and decreased *N*-acetyl aspartate/creatine (NAA/Cr) ratios within the lesion.
   - These findings are consistent with low-grade gliomas or demyelination processes.[1,2]
3. *How can you anatomically explain the CN VI and VII palsies?*
   - This lesion involves the facial colliculus, which is formed by the facial motor fibers as they circle around the abducens nucleus in the dorsum of the pons.[3]
   - A lesion in the facial colliculus affects both the facial motor fibers and the abducens nucleus (▶**Fig. 67.4**).
4. *If you chose to operate, what would be the aim of the surgery?*
   - The aim of surgery is two-fold:
     - Decompression of the pons
     - Obtaining tissue for diagnosis
5. *How would you approach this lesion?*
   - Suboccipital craniotomy–vermis-sparing telovelar approach
   - The infrafacial triangle may be utilized for approaching the tumor. As this lesion is occupying the facial colliculus and is pointing to the floor of the fourth ventricle, this lesion should be approached through the infrafacial triangle (that can be found by mapping or measurements) and/or the area closest to the surface of the fourth ventricle (▶**Fig. 67.4**).[4–6]
   - This approach would be further evaluated by use of neuronavigation and microscopy (▶**Fig. 67.5**).
   - In this particular case, an 8-mm area from the presumed midline and just below the striae medullares was used as the center of the infrafacial triangle and the closest part of the tumor to the fourth ventricle floor.
6. *What neurophysiologic modalities would you utilize during surgery?*
   - Three modalities are available to monitor this patient[7,8] (electrodes were checked and tested on a volunteer for MRI compatibility and the patient consented to monitoring):
     - Brainstem auditory evoked responses
     - Sensory evoked potentials
     - Motor evoked potentials
     - Fourth ventricular floor mapping
7. *What are the safe entry zones into the floor of the fourth ventricle?*
   - Safe entry zones into the floor of the fourth ventricle include (▶**Fig. 67.4**):
     - Suprafacial (supra-abducental) triangle: a triangle measuring around 16 mm in longest diameter. It is located above the facial colliculus and 5 mm from the midline (to avoid the medial longitudinal fascicle [MLF]). Its upper and narrower angle is below the trochlear nucleus.[5,6]
     - Infrafacial (infra-abducental) triangle: a smaller triangle measuring less than 9 mm located just below the facial colliculus. It is narrow as it is located between the MLF medially and the facial nucleus laterally.[5,6]
8. *How would you further manage this case?*
   - Management plan includes:
     - For the residual tumor: conformal radiation or gamma knife treatment to the pons can be given postoperatively. This can be followed by serial MRI and close follow-ups.[9,10]
     - Note that, however, due to the relative risk and safety issues involved in using gamma knife treatments to the pons, some would not consider this option unless the lesion was exophytic from the pons.
     - For the facial palsy: teardrops and eye protection. One may resort to partial or gold-weight tarsorrhaphy in some cases to avoid corneal abrasions and ulcers. Facial nerve reanimation procedures can also be tried.
     - For the gait disturbances: continued inpatient or outpatient rehabilitation
9. *Classify pontine tumors.*
   - Classification of pontine tumors: pontine tumors can be classified on the basis of type of tumor or growth pattern[11]:
     - Diffused pontine tumors: usually malignant and difficult to delineate from neighboring pontine parenchyma. Usually low or isointense signal on T1-weighted MRI and may have an increased signal on T2-weighted MRI. Hyperintensity on T1-weighted MRI is usually due to hemorrhagic change. Further enhancement in these diffuse tumors may be a sign of actual malignant degeneration.
     - Focal pontine tumors: well-demarcated and hypo- or isointense signal on T1-weighted and high signal intensity on T2-weighted MRIs.
     - Exophytic pontine tumors: almost always dorsally exophytic into the fourth ventricle and may be benign or malignant.[12,13]

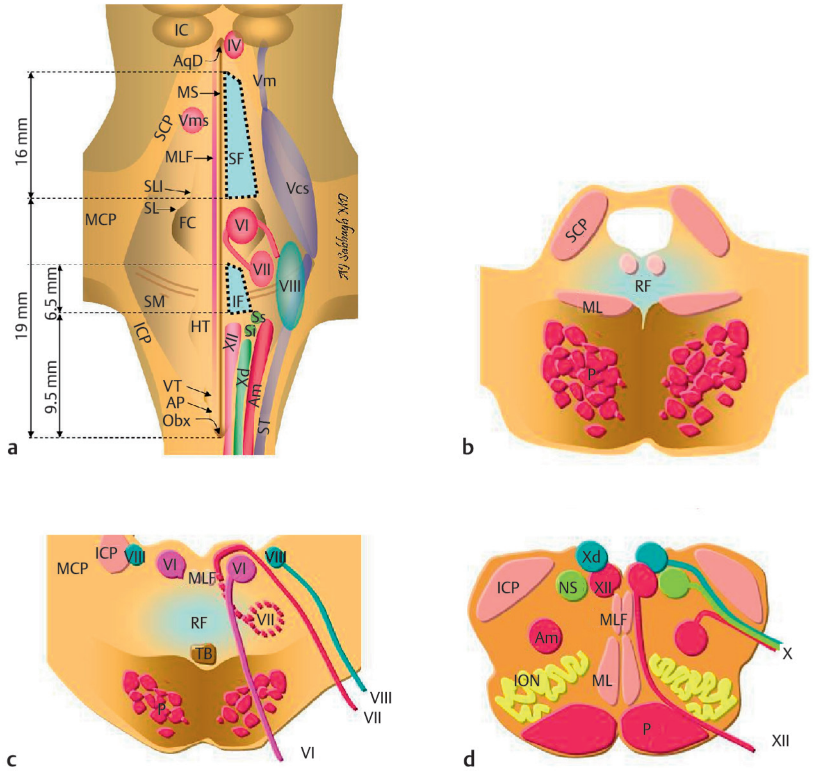
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**Fig. 67.4** Safe entry zones of the brainstem. Artist's rendering of the brainstem from a dorsal view (**a**) with illustrated safe entry zones and relevant nuclei and neural structures. The infrafacial and suprafacial triangles are highlighted as safe entry zones in the dorsal pons. Corresponding axial sections through the (**b**) upper, (**c**) mid, and (**d**) lower pons are illustrated. AqD, aqueduct of Sylvius; N, nucleus; IC, inferior colliculus; MS, median sulcus; Vm, mesencephalic N. of the 5th cranial nerve (V); Vcs, chief (sensory) N. of V; Vms, motor (mastication) N. of V; MLF, medial longitudinal fascicle; FC, facial colliculus; IV, trochlear N.; CTT, central tegmental tract; SL, sulcus limitans; SLI, sulcus limitans incisure; HT, hypoglossal triangle; SM, striae medullares; SCP, MCP, ICP, superior, middle, and inferior cerebellar peduncle; VT, vagal triangle; AP, area postrema; Obx, Obex; VI, abducent N.; VII, facial N. and fiber tracks and nerve; VIII, vestibular N. and nerve; XII, hypoglossal N. and nerve; Xd, dorsal vagal N.; Am, N. ambiguus of ninth and tenth cranial nerves with parasympathetics on its medial border; Ss & Si, superior and inferior salivatory NN.; ST, spinal trigeminal tract; STT, spinothalamic tract; ML, medial lemniscus; ION, inferior olivary N.; P, pyramid; TB, trapezoid body; Pn TPF, pontine NN and transverse pontine fibers; SF, suprafacial triangle; IF, infrafacial triangle.

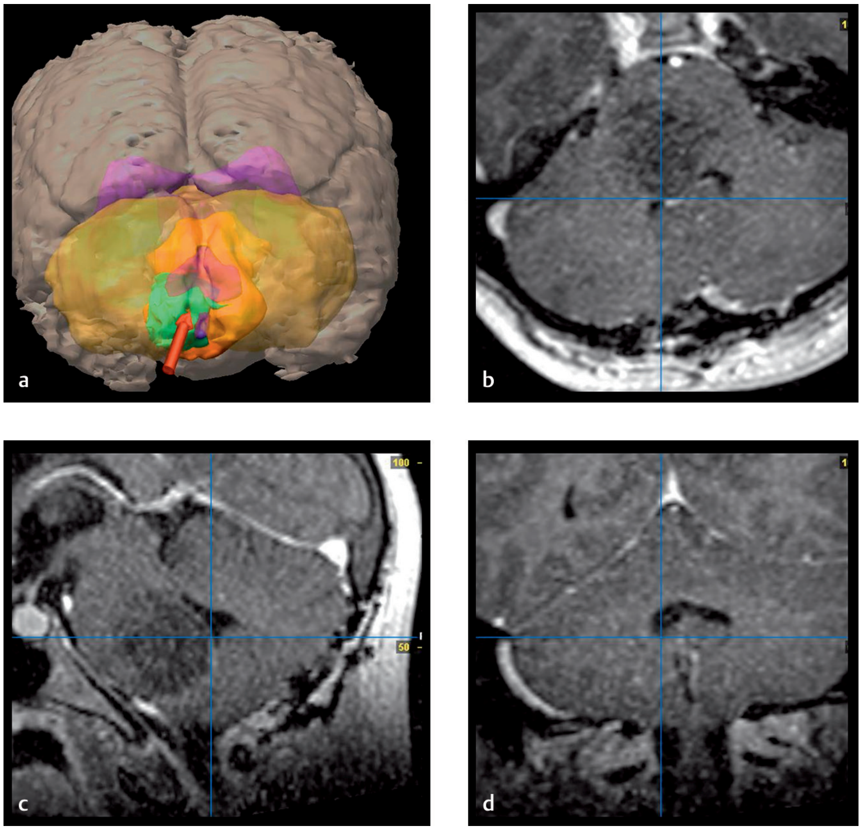
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**Fig. 67.5** Intraoperative neuronavigation image with three-dimensional reconstruction (**a**) highlighting tumor in *green* and entry trajectory with *red arrow*. Corresponding T1-weighted MR images with contrast with (**b**) axial, (**c**) sagittal, and (**d**) coronal views.

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# Case 68 Pineal Region Tumors

*Kathleen E. Knudson, John S. Myseros, and Robert F. Keating*

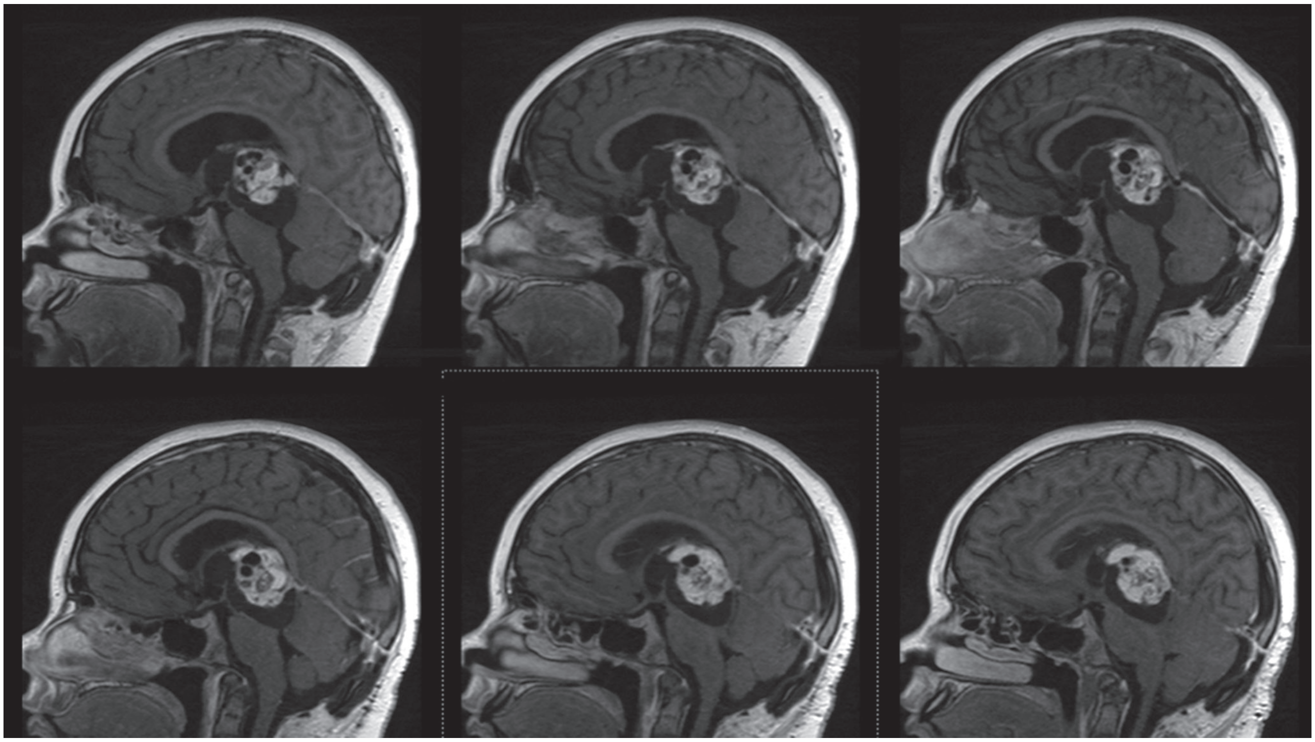

**Fig. 68.1** MRI brain midsagittal T1 with contrast; pathology consistent with mature teratoma.

## Clinical Presentation

- A 7-year-old male presents with the complaints of headache, vomiting, and double vision.
- The patient's neurological exam is significant for papilledema, paralysis of upward gaze, and mild ataxia.
- The patient's MRI brain mid-sagittal T1-weighted images with contrast are shown in ▶**Fig. 68.1.**

## Questions

1. What is the clinical presentation of a patient with a pineal region tumor?
2. What is the differential diagnosis of a lesion in the pineal region?
3. What are different characteristics of pineal region tumors on imaging?
4. What additional work-up should be performed for a patient with a pineal region lesion?
5. What are the different surgical approaches to the pineal region?
6. What are the adjuvant treatments for pineal region tumors?

## Answers

1. *What is the clinical presentation of a patient with a pineal region tumor?*
    - Symptoms related to increased intracranial pressure from hydrocephalus
        - Headaches, nausea, vomiting, obtundation, and papilledema
    - Symptoms related to brainstem compression
        - Parinaud's syndrome: compression of the superior colliculus can result in paralysis of upward gaze, convergence or retraction nystagmus,
          and near-light pupillary dissociation.[1]
        - Sylvian aqueduct syndrome: paralysis of downward gaze or horizontal gaze[2]
    - Symptoms related to cerebellar compression
        - Ataxia, dysmetria
    - Endocrine dysfunction
        - Due to hydrocephalus or spread to the hypothalamus
        - Diabetes insipidus, precocious puberty[3]
2. *What is the differential diagnosis of a lesion in the pineal region?*
    - Pineal parenchymal neoplasms: lesion arising from the cells of the pineal gland
        - Pineocytoma: low-grade lesion, WHO grade I
            - ○ Usually occurs around 40 years of age
            - ○ Usually compress surrounding tissues instead of infiltrating
        - Intermediate pineal parenchymal tumor: WHO grade II, III
        - Pineoblastoma: high-grade lesion, WHO grade IV
            - ○ Usually occurs at 20 years of age
            - ○ Tendency to infiltrate surrounding tissues and disseminate through the central nervous system (CNS) along cerebrospinal fluid (CSF) pathways
    - Germ cell tumors (GCTs): derive from pluripotential germ cells
    - Germinoma: most common subtype
            - ○ Well-circumscribed GCT usually occurring in the pineal and suprasellar regions
        - Nongerminomatous GCTs
            - ○ Embryonal carcinoma
            - ○ Yolk sac tumors
            - ○ Choriocarcinoma: contains syncytiotrophoblastic cells, commonly associated with hemorrhage
            - ○ Teratoma: tumor that contains a mixture of tissues derived from the three embryonic cell lines
            - ○ Mixed GCT: encompasses a combination of any of the GCTs
    - Gliomas: tend to be infiltrating
    - Others:
        - Meningioma, ependymoma
        - Melanoma
        - Metastatic tumors
        - Cysts
    - Vascular lesions
        - Cavernous malformation, arteriovenous malformation, vein of Galen malformation
3. *What are different characteristics of pineal region tumors on imaging?*
    - MRI brain with contrast is the diagnostic imaging study of choice for pineal region neoplasms.
    - MRI can give important information about the tumor size, composition, and relationship with surrounding structures.[4]
    - However, MRI cannot reliably determine tumor pathology.
    - Some imaging findings can be more characteristic of certain tumor pathologies.
        - Germinomas tend to be more homogenously enhancing when compared to nongerminomatous germ cell tumors.
        - Teratomas contain elements from multiple germ cell layers that can often be identified in MRI.[5]
    - ▶ **Fig. 68.1** is the MRI from our patient—the pathology was consistent with mature teratoma.
4. *What additional work-up should be performed for a patient with a pineal region lesion?*
    - CSF markers are more sensitive than serum marker levels[6]—the three main markers are placental alkaline phosphatase (PALP), alpha fetoprotein (AFP), and human chorionic gonadotropin (hCG).
    - Patients with a germinoma tend to have elevated levels of PALP, without elevation in AFP or hCG.
        - If AFP or hCG are elevated, this could indicate that the tumor is a mixed GCT.
    - Yolk sac tumors will show elevated levels of AFP.
    - Choriocarcinoma will show elevated levels of hCG.
    - Pineocytomas/pineoblastomas will not have elevations in any of these tumor markers.
    - Teratomas, embryonal carcinomas, and mixed GCTs will have a mixture of these hormones.[7]
    - Tumor markers can also be used to monitor progress and response to treatment.
5. *What are the different surgical approaches to the pineal region?*
    - Surgical options include stereotactic biopsy versus endoscopic biopsy versus open resection.
        - Stereotactic biopsy:
            - ○ Benefits: minimally invasive, quick recovery
            - ○ Risks: insufficient tissue for diagnosis, tissue not representative of only one portion of a mixed GCT, bleeding or vascular (internal cerebral vein [ICV]) injury, tumor bleeding[8,9]
        - Endoscopic biopsy:
            - ○ Benefits: minimally invasive, concomitant treatment of hydrocephalus with endoscopic third ventriculostomy (ETV), direct visualization of

## Answers (*continued*)

    - tumor, irrigation available to stop any potential bleeding, potential biopsy of various areas of the tumor
    - Risks: uncontrollable bleeding, third ventricular/hypothalamic injury, forniceal injury, insufficient tumor sampling[10,11]
  - Open resection:
    - Benefits: optimal visualization and exposure of tumor, maximal safe resection, control of bleeding, potential of cure of hydrocephalus with adequate resection, sampling of various areas of the tumor
    - Risks: general craniotomy risks, bleeding, dural venous sinus injury, deep venous injury, midbrain/thalamic injury[8]
- Open surgical approaches:
  - Infratentorial supracerebellar approach:[12,13]
    - Patient positioned in a concord or "park bench" position with midline incision extending from above the inion to the C2 spinous process
    - Perform the craniotomy over the sagittal sinus and both lateral sinuses about 1 to 2 cm above foramen magnum.
    - Open the dura in a semilunar curve that is reflected upward to avoid retraction of sinus.
    - Open the infratentorial corridor with careful dissection of any veins especially the precentral vermian vein, with a wide opening of the arachnoid.
    - Microscopic resection of the tumor: internal debulking followed by dissection of the capsule from surrounding tissues
    - Once tumor resection is complete, one should be able to see into the third ventricle.
    - Advantages to this approach: following a natural corridor between the tentorium and cerebellum[13] and visualization of the vein of Galen and the basal vein of Rosenthal (BVR)
    - Disadvantages/limitations: small working corridor at the incisura, hemorrhage can occur above tumor and may be difficult to visualize
  - Transcallosal interhemispheric approach:
    - Patient positioned prone, head in neutral position
    - Wide craniotomy with burr holes over the sagittal sinus
    - Follow down corridor along parieto-occipital junction between the falx and the hemisphere
    - Identify the corpus callosum and make a 1 to 2 cm callosotomy
    - Open tela choroidea and arachnoid over the internal cerebral vein (ICV)
    - Tumor resection is performed between the ICVs
    - Tentorium/falx can be divided to provide additional exposure.
    - Proceed with tumor debulking and resection.
    - Advantages of this approach: good for tumors that extend superiorly/supratentorially[8] (e.g., into the third ventricle)
    - Disadvantages/limitations: limited by working between the veins (ICVs), BVR not immediately visualized
    - Intraoperative image from interhemispheric transcallosal approach to resection of this pineal tumor; the tumor is exposed and resected between the two ICVs (▶**Fig. 68.2**).
  - Occipital transtentorial approach:
    - Similar approach as the transcallosal interhemispheric but with a more oblique trajectory
    - Retract the nondominant occipital lobe and divide the tentorium.
    - Proceed with tumor debulking and resection.
    - Advantages of this approach: direct visualization of veins, access to vermis/posterior fossa
    - Disadvantages/limitations: retraction of occipital lobe, poor visualization of posterior third ventricle

6. *What are the adjuvant treatments for pineal region tumors?*
   - Adjuvant treatments are usually determined based on tumor pathology, as this will determine their sensitivity to treatment.
   - Radiation therapy:
     - Radiation is typically performed to the tumor bed/pineal region and ventricular system.
     - Germinomas are very sensitive to radiation with 5-year survival rates > 75%.
     - Nongerminomatous GCTs are less sensitive, with 5-year survival rates of about 30 to 40%.
     - Radiation can be withheld for low-grade pineocytomas or ependymomas that have been completely resected.
     - Complications of radiation include: cognitive deficits, hypothalamic/endocrine dysfunction, cerebral necrosis, de novo tumor formation
   - Chemotherapy:
     - GCTs are generally more sensitive to chemotherapy than pineal cell tumors.
     - Typical regimen includes cisplatin/carboplatin and etoposide.
       - Most treatment regimens are extrapolated from the treatment of extracranial tumors.
   - The best survival rates in cases of aggressive tumors necessitate a combination of chemotherapy, radiation, and surgery.

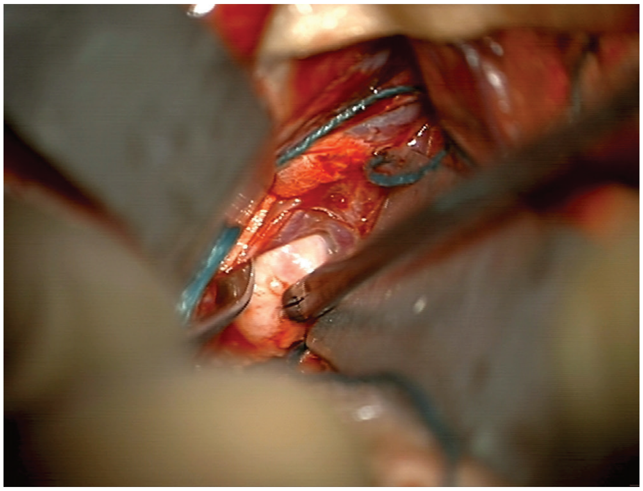

**Fig. 68.2** Intraoperative image from interhemispheric transcallosal approach to resection of pineal tumor, the tumor is exposed and resected between the two internal cerebral veins.

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# Case 69 Posterior Fossa Ependymoma

*Philippe Mercier and Frederick Boop*

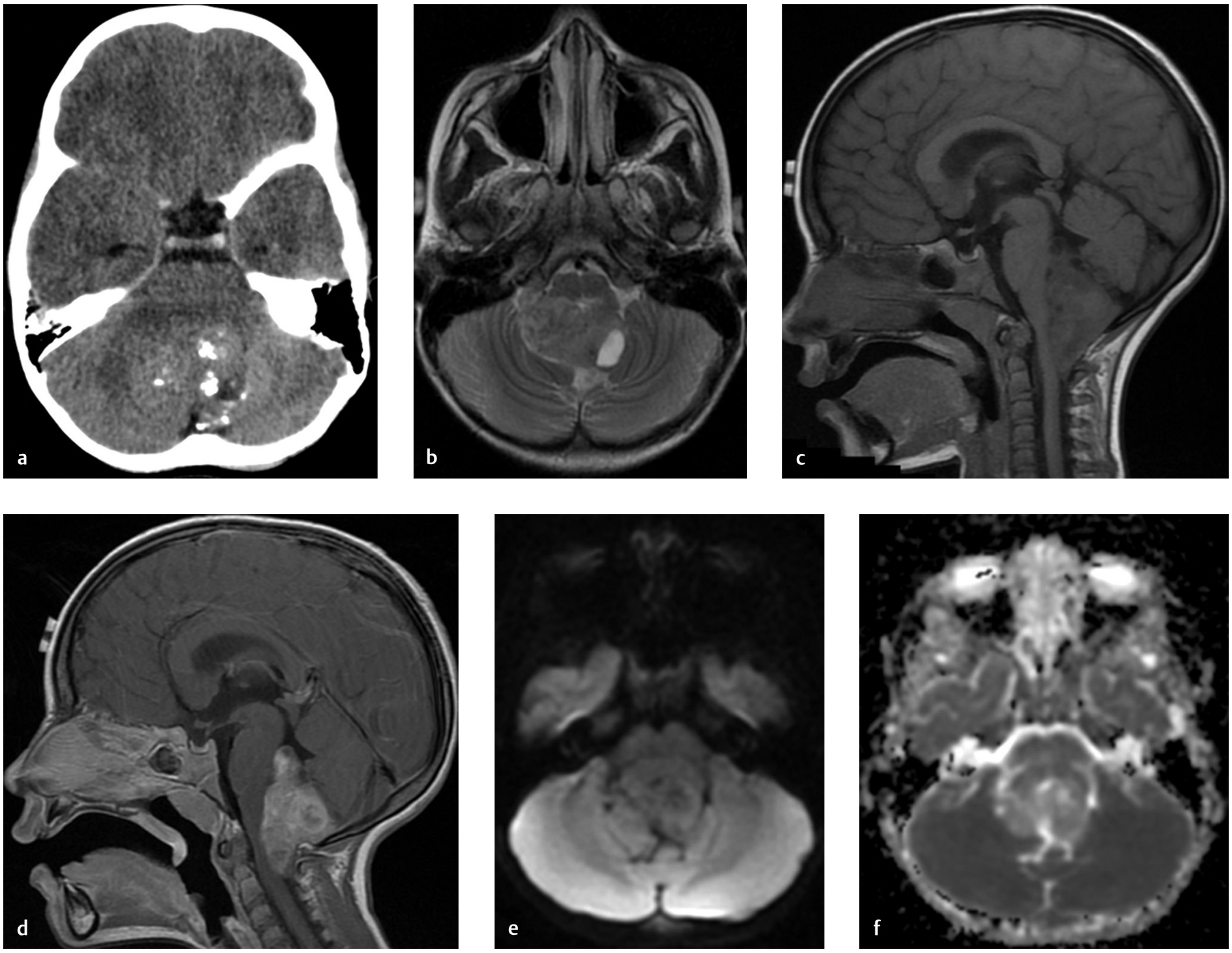

**Fig. 69.1** Axial CT of the head demonstrates a mass in the posterior fossa with internal calcifications resulting in narrowing of the fourth ventricle (**a**). The lesion is heterogenous on axial T2-weighted MRI of the brain (**b**) with extension of the lesion into the foramen of Luschka. The lesion is solidly enhancing comparing sagittal T1-weighted (**c**) and T1-weighted with contrast images (**d**) extending from the floor of the fourth ventricle into the upper cervical spine. The lesion on diffusion-weighted imaging is dark/hypointense (**e**) and on apparent diffusion coefficient (**f**) is bright/hyperintense suggesting a lack of diffusion restriction.

## Clinical Presentation

- A 5-year-old female presents to the hospital with a 4-month history of nausea, vomiting, and weight loss and a several-week history of headache and neck pain.
- On examination, she is awake, alert, answering questions appropriately for her age, and her pupils are equal and reactive to light; she has a symmetrical face, loss of gag; she has good motor strength throughout all extremities and sensory examination is intact to light touch with mild 3+ hyperreflexia at the patellae (▶**Fig. 69.1**).

## Questions

1. What is the differential diagnosis?
2. What features of this imaging suggest the diagnosis of ependymoma?
3. What additional testing should be performed to assess a child with posterior fossa ependymoma?
4. How are intracranial ependymomas classified pathologically?
5. Describe the role of surgical management in the treatment of ependymoma.
6. What surgical approaches can be used for resection of this tumor?
7. What is the role of radiation in the management of posterior fossa ependymoma?
8. Is adjuvant therapy recommended in the treatment of ependymoma?
9. How often is ventriculoperitoneal shunt required in the treatment of hydrocephalus associated with ependymoma?
10. What determines prognosis in ependymoma?

## Answers

1. *What is the differential diagnosis?*
   - Ependymoma
   - Medulloblastoma
   - Pilocytic astrocytoma
   - Atypical teratoid/rhabdoid tumor
   - Ganglioglioma
   - Metastasis (rare in children)
2. *What features of this imaging suggest the diagnosis of ependymoma?*
   - On CT of the brain, ependymoma is typically contained within the fourth ventricle and extends to the foramen of Luschka and Magendie; it is hypodense and can be cystic with coarse calcifications.
   - With MRI of the brain, ependymoma is typically hypointense on T1-weighted images, hyperintense on T2-weighted images, and displays heterogeneous enhancement. Ependymoma can restrict diffusion but not as often as the more cellular medulloblastoma.[1]
   - The incidence of spinal metastases upon presentation of ependymoma is 5%.[2]
3. *What additional testing should be performed to assess a child with posterior fossa ependymoma?*
   - MRI of the spinal axis with and without contrast is used to assess for the presence of drop metastases that can coat the dura and nerve roots.
   - In addition, lumbar puncture is typically performed 2 to 4 weeks postoperatively and sent for cytology to monitor for metastatic dissemination.
4. *How are intracranial ependymomas classified pathologically?*
   - Intracranial ependymoma have several variants including subependymoma (WHO grade I) that occur in adults and very rare intracranial myxopapillary ependymoma.
   - The majority of intracranial ependymomas occurring in children are graded as WHO grade II and can have cellular, papillary, clear cell, and tanycytic histological subtypes.
   - Anaplastic (WHO grade III) ependymomas that typically have pleomorphism, are multinucleated, can have giant cells, mitotic figures, vascular changes, and areas of necrosis.
   - A recent review of a series of grade II and III ependymomas revealed the difficulties in assigning specific grades to different tumors based on histology, resulting in variability in the diagnosis and prognosis.[3]
   - A specific molecular subtype, RELA-fusion positive, can be described for supratentorial ependymoma, but there are no molecular markers that can reliably aid in differentiating grade II and III ependymomas that are described in the latest WHO classification of central nervous system tumors.[4]
5. *Describe the role of surgical management in the treatment of ependymoma.*
   - The surgical management of posterior fossa ependymoma consists of maximal safe surgical resection regardless of WHO grade (II or III).[5]
6. *What surgical approaches can be used for resection of this tumor?*
   - Preoperative treatment of children with these lesions should include corticosteroids.
   - In addition, an external ventricular drain can be used to treat hydrocephalus and aid in safer dissection of the lesion during resection.
   - Typically, these lesions can be removed using either transvermian, transcortical, or telovelar approaches.[6]
7. *What is the role of radiation in the management of posterior fossa ependymoma?*
   - Conformal photon therapy is typically delivered to the operative bed after surgical resection at 54 to 59.4 Gy with a 1.0 cm margin.
   - If there are spinal metastases on presentation or if CSF cytology is positive, low dose radiotherapy is delivered to the spine.[5]
   - Proton radiotherapy is being explored as a means to spare structures around the tumor margin.

## Answers (*continued*)

8. *Is adjuvant therapy recommended in the treatment of ependymoma?*
   - The role of chemotherapy remains controversial and is typically reserved for young children under 3 in order to delay the use of radiation.[5]
9. *How often is ventriculoperitoneal shunt required in the treatment of hydrocephalus associated with ependymoma?*
   - Approximately 70 to 90% of patients with posterior fossa tumors present with hydrocephalus.
   - The Canadian Preoperative Prediction Rule for Hydrocephalus identifies the following risk factors that increase in the chance of a patient having persistent hydrocephalus post resection[7]:
     - Age less than 2
     - Preoperative papilledema
     - Moderate to severe hydrocephalus upon presentation
     - Cerebral metastases
     - Medulloblastoma
     - Ependymoma
     - Dorsally exophytic glioma
10. *What determines prognosis in ependymoma?*
    - The most significant prognostic variable in children with posterior fossa ependymoma is extent of resection.[8]
    - Although not named in the WHO classification of ependymoma, there is evidence for two distinct molecular subgroups of ependymoma in the posterior fossa designated EPN-A and EPN-B.[9]
      - EPN-A occurs primarily in young children carrying a worse prognosis.
      - EPN-B occurs in older children and adults and carries a better prognosis regardless of WHO histologic diagnosis.[8]
    - In addition, the most frequent copy number alteration in ependymoma, 1q gain, is predictive of poor outcome and is present across both EPN-A and EPN-B subtypes.[10,11]

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# Case 70 Neurofibromatosis Type 1

*Jean-Pierre Farmer and Abdulrahman J. Sabbagh*

## Clinical Presentation

- A 3-year-old child is referred to you because of the presence of multiple café-au-lait spots.
- His parents have a normal phenotype.
- History does not reveal any developmental delay.
- His physical examination shows macrocrania, multiple café-au-lait spots, 12 in total, greater than 0.5 cm.
- There is no scoliosis.
- He has pulsatile proptosis of the right eye without chemosis or pupillary changes.

## Questions

1. What are the diagnostic criteria for neurofibromatosis type I (NF-1)?
2. What are other NF-1-associated conditions?
3. What are the types of cutaneous neurofibromas?
4. How would you proceed with investigations?
5. Which additional consultants would you involve?
6. Which lesions, if present, would likely require more frequent surveillance and imaging in this patient?
7. What is the cause of pulsatile proptosis?
8. What is the chance of occurrence of optic pathway gliomas in NF patients?
9. What are the rates of optic glioma progression in NF patients?
10. What imaging characteristics of optic glioma might help in determining the prognosis?
11. What is the usual cause of aqueductal stenosis in NF-1 patients?
12. How frequent are academic difficulties in patients with NF?
13. Assuming the MRI of the brain and spine are normal, what would be your recommended follow-up scan frequency?
14. Is the age at diagnosis significant with respect to prognosis in patients with NF-1?
15. What is the NF-1 gene and where is it located?
16. What is neurofibromin? What are its functions?
17. Describe the Ras signaling pathway (Rat sarcoma) and its relation to neurofibromin.

## Answers

1. *What are the diagnostic criteria for neurofibromatosis type I (NF-1)?*
   - NF-1 criteria include the following:[1]
     - Six or more café-au-lait spots (over 5 mm in prepubertal individuals, over 15 mm in postpubertal individuals)
     - Two or more neurofibromas of any type or one plexiform neurofibroma
     - Freckling in the axillary or inguinal region
     - Optic glioma
     - Two or more Lisch nodules (iris hamartomas)
     - Osseous lesions such as sphenoid dysplasia or thinning of the long bone cortex with or without pseudarthrosis
     - A first-degree relative (parent, sibling, or off spring) with NF-1 by the above criteria
2. *What are other NF-1-associated conditions?*
   - NF-1-associated conditions include the following:[1,2]
     - Aqueductal stenosis
     - Macrocephaly
     - Unilateral superior orbital defect (pulsatile exophthalmos)
     - Cognitive impairment and learning disabilities
     - Kyphoscoliosis
     - Syringomyelia
     - Intracranial tumors
       - Astrocytoma, hemispheric
       - Meningioma, solitary or multicentric (adults)
     - Extracranial tumors
       - Schwann cell tumors
       - Neuroblastoma
       - Sarcoma
       - Leukemia
       - Wilms' tumor
       - Pheochromocytoma
     - Moyamoya disease
3. *What are the types of cutaneous neurofibromas?*
   - Types of neurofibromas (Friedman and Riccardi classification):[3–5]

## Answers (*continued*)

  - Discrete cutaneous: Soft and fleshy nodules located mainly in epidermis, dermis, and trunk, and in the face and extremities as well. Usually more than 100 lesions are seen by 40 years of age.
  - Discrete subcutaneous: These are firm and rubbery.
  - Deep nodular (plexiform nodular): Involve nerves beneath the dermis; they are fusiform; may involve the entire nerve.
  - Diffuse plexiform: congenital in origin, evident in infancy or childhood. They are locally invasive and have poorly defined margins. They may be precancerous.

4. *How would you proceed with investigations?*
   - MRI of the brain, spine, and orbits
   - Formal visual acuity, visual fields, and extraocular movement testing
   - Genetic testing
5. *Which additional consultants would you involve?*
   - Ophthalmologist
   - Pediatrician
   - Geneticist
6. *Which lesions, if present, would likely require more frequent surveillance and imaging in this patient?*
   - Lesions requiring frequent follow-ups include:[6,7]
     - Brainstem gliomas
     - Paraspinal gliomas
     - Craniofacial neurofibromas
     - Symptomatic lesions
     - Extraoptic glioma
   - Every 2 years for asymptomatic gadolinium-enhancing lesions[7]
     - Optic pathway and parenchymal gliomas
     - Cranial nerve and visceral neurofibromas
7. *What is the cause of pulsatile proptosis?*
   - The most likely cause of pulsatile proptosis is sphenoid wing hypoplasia.
8. *What is the chance of occurrence of optic pathway gliomas in NF patients?*
   - The chance of occurrence of optic pathway gliomas in NF patients is 15 to 30%.[7,8]
9. *What are the rates of optic glioma progression in NF patients?*
   - Optic pathway gliomas progress in 12% of NF patients who harbor them.
10. *What imaging characteristics of optic glioma might help in determining the prognosis?*
    - The main characteristic that indicates a smaller chance of progression is lack of enhancement.[7]
11. *What is the usual cause of aqueductal stenosis in NF-1 patients?*
    - The usual cause of aqueductal stenosis in NF-1 patients is the occurrence of midbrain unidentified bright objects (UBOs).[9,10]
12. *How frequent are academic difficulties in patients with NF?*
    - Academic difficulties in patients with NF-1 are as common as 30 to 46%.[11,12]
13. *Assuming the MRI of the brain and spine are normal, what would be your recommended follow-up scan frequency?*
    - Assuming the MRI of the brain and spine are normal, young patients require annual MRIs.[13]
14. *Is the age at diagnosis significant with respect to prognosis in patients with NF-1?*
    - Age at diagnosis below 6 years carries a significantly worse prognosis in patients with NF-1.
15. *What is the NF-1 gene and where is it located?*
    - The NF-1 gene has the following characteristics:[12,14]
      - Located on chromosome 17, band q12.2
      - Base pairs 26, 446, 242 to 26, 725, 589
      - It has 335,000 chemical bases
      - Sixty exons, alternative splicing at exons 9a, 23a, 48a
      - It is a tumor suppressor gene that encodes neurofibromin.
16. *What is neurofibromin? What are its functions?*
    - Neurofibromin[15]
      - It is a protein (2818 amino acids, molecular mass 327 kDa)[16] that is mainly expressed in:
        - Astrocytes and oligodendrocytes of the central nervous system
        - Sensory neurons of the peripheral nervous system
        - Schwann cells
        - Other cells originating from the neural crest such as melanocytes
      - It functions by inactivating Ras by stimulating intrinsic Ras-GTPase to hydrolyze Ras attached guanosine triphosphate (GTP) to guanosine diphosphate (GDP).
17. *Describe the RAS signaling pathway (Rat sarcoma) and its relation to neurofibromin.*
    - RAS when activated can lead to tumor formation.
    - Neurofibromin functions to deactivate RAS.
    - See ▶ **Fig. 70.1** for further details.

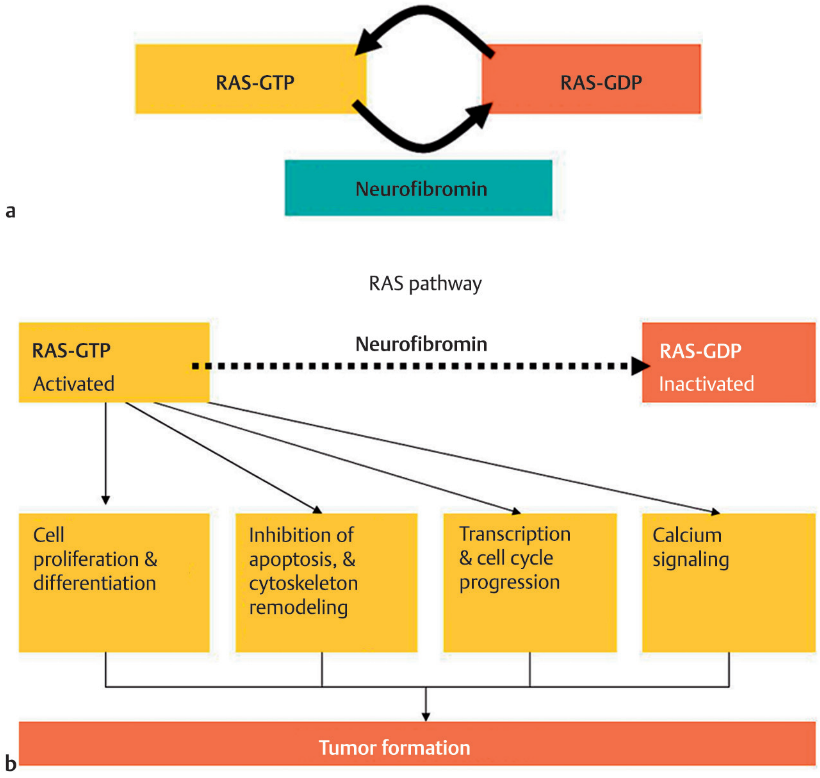
RAS-GTP
RAS-GDP
Neurofibromin
a
RAS pathway
RAS-GTP
Activated
Neurofibromin
RAS-GDP
Inactivated
Cell proliferation & differentiation
Inhibition of apoptosis, & cytoskeleton remodeling
Transcription & cell cycle progression
Calcium signaling
Tumor formation
b

**Fig. 70.1** Illustration of molecular genetics of Ras pathways: (**a**) Ras activation and deactivation, and (**b**) Ras pathway for tumor formation.

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# Case 71 Hypothalamic Hamartoma

*Abdulrahman J. Sabbagh, Sandeep Mittal, Fahad Eid Alotaibi, and José Luis Montes*

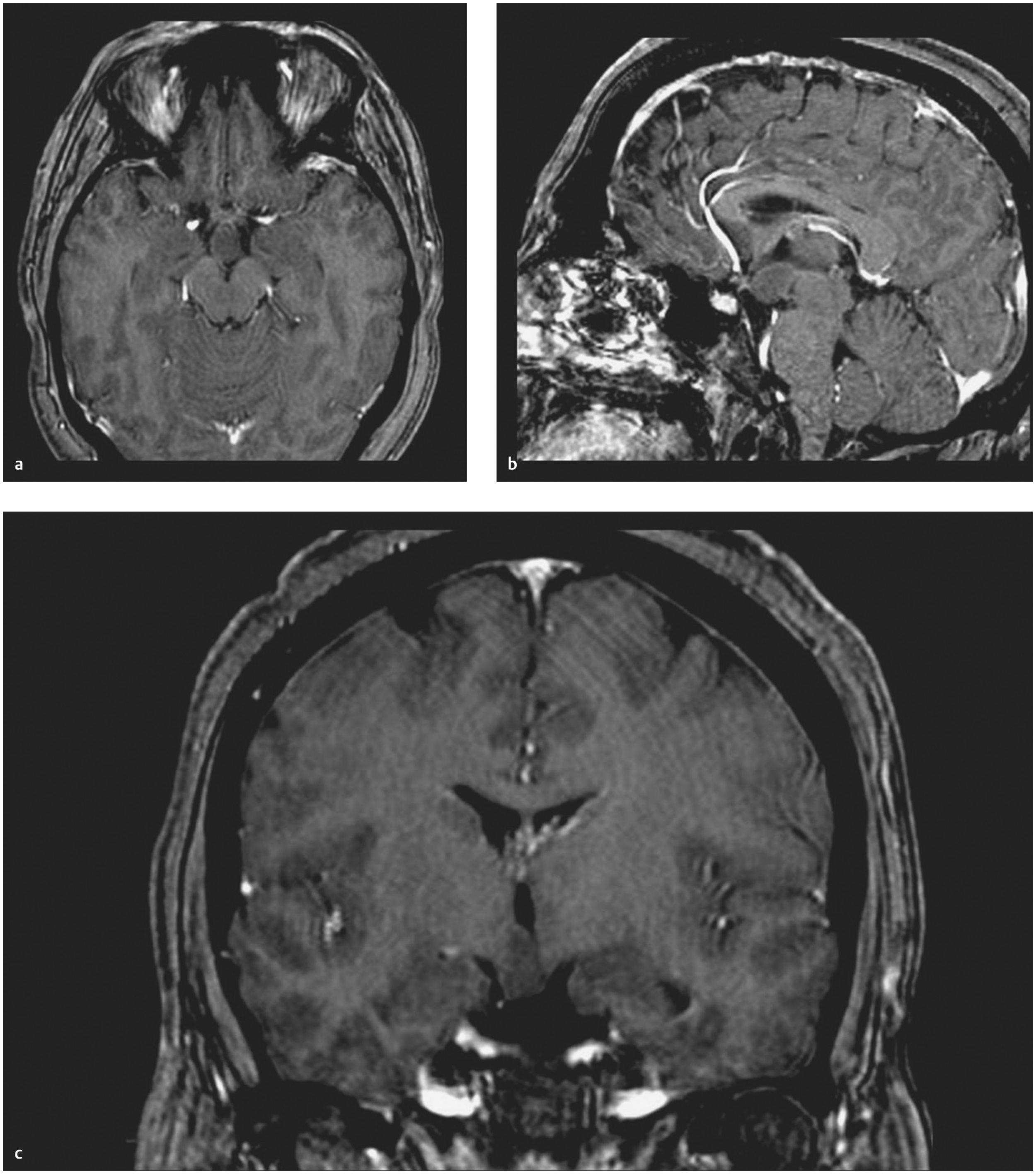


**Fig. 71.1** T1-weighted MR images of the brain with contrast, relevant (**a**) axial, (**b**) sagittal, and (**c**) coronal slices are shown.

## Clinical Presentation

- A 16-year-old boy is referred to you by an epileptologist. He presents with a history of progressive epilepsy that is refractory to medication.
- The seizures are described as episodes of short bouts of emotionless laughter with loss of awareness that last only a few seconds at a time and occur several times during the day now.
- He also suffers from generalized tonic–clonic seizures several times a month.
- Other pertinent findings include cognitive delay. He is able to speak. The remainder of his neurologic exam is normal (including motor, sensory, cerebellar, and gait examination).
- An MRI scan is obtained and shown in ▶**Fig. 71.1**.

## Questions

1. Describe the MRI (▶**Fig. 71.1**).
2. What is the term used for type of seizures this patient is experiencing?
3. How will you work up this patient?
4. Briefly describe the anatomy of the hypothalamus; enumerate its nuclei and their functions.
5. What seizure types are associated with hypothalamic hamartomas (HH)?
6. What symptoms other than epilepsy are associated with HH?
7. If this patient has polydactyly and hypopituitarism, what syndrome would you want to confirm?
8. How do you classify HH? What class does this patient's hamartoma belong to?
9. What treatment options can you offer to a patient with HH and intractable seizures? What are their limitations and outcomes?

## Answers

1. *Describe the MRI* (▶**Fig. 71.1**).
    - MRI shows a small nonenhancing, isointense lesion in the hypothalamic area.
    - The mass occupies the third ventricle and does not cause hydrocephalus.
    - This is most consistent with an HH.
2. *What is the term used for type of seizures this patient is experiencing?*
    - Gelastic seizures
3. *How will you work up this patient?*
    - Imaging and electrophysiologic investigations include:
        - Electroencephalogram (EEG): can show slow spike and wave EEG patterns with or without multifocal epileptiform abnormalities (typically frontal or temporal)[1]
        - CT scan: may show an isodense nonenhancing lesion
        - MRI: shows an isointense to slightly hypointense lesion compared with gray matter[2]
        - Depth electrode recording: When the diagnosis is equivocal and EEG is nonspecific, specialized centers may consider this modality for diagnosis (▶**Fig. 71.2**).[3,4]
        - Positron emission tomography scan: reveals ictal hypermetabolism at the hamartoma site[5]
        - Single photon emission CT (SPECT) imaging: measures regional cerebral blood flow during seizures. Ictal SPECT scans can be done after injection of the tracer technetium-99m hexamethylpropyleneamine oxime (Tc-99m-HMPAO).
        - Magnetoencephalography (MEG): MEG maps interictal magnetic dipole sources onto MRI to produce a magnetic source image.[6]
        - MR spectroscopy: decrease in *N*-acetyl aspartate/creatine and an increase in myoinositol/creatine (mI/Cr) ratios in tumor tissue when compared with values in normal gray matter of the amygdala. Choline/creatine ratios were also increased when compared with those in normal gray matter controls.[7]
    - Endocrinologic work-up
        - See Case 12, Pituitary Adenoma, for details.
4. *Briefly describe the anatomy of the hypothalamus; enumerate its nuclei and their functions.*
    - The hypothalamus is commonly subdivided into regions along its anteroposterior axis (▶**Fig. 71.3**).[8]
        - The preoptic region extends rostrally to the optic chiasm and dorsally to the anterior commissure.
        - The supraoptic region resides above the optic chiasm.
        - The tuberal region lies above and includes the tuber cinereum.
        - The mammillary region includes the mammillary bodies and the posterior hypothalamic nuclei.
5. *What seizure types are associated with hypothalamic hamartomas (HH)?*
    - Seizure types include:
        - Gelastic seizures (forced bouts of emotionless laughter) are considered by most authors to be characteristic of HH.

## Answers (*continued*)

  - Multiple other seizure types include:
    - Generalized tonic–clonic seizures
    - Partial complex seizures
    - Drop attacks
    - Atypical absences

6. What symptoms other than epilepsy are associated with HH?
   - Other associated symptoms may include:
     - Precocious puberty
     - Psychiatric manifestations such as:[9,10]
       - Oppositional defiant disorder (83.3%)
       - Attention deficit hyperactivity disorder (75%)
       - Conduct disorder (33.3%)
       - Affective disorders (16.7%)
       - Progressive cognitive decline
7. *If this patient has polydactyly and hypopituitarism, what syndrome would you want to confirm?*
   - Pallister–Hall syndrome (PHS)[11]
   - The syndrome is typically characterized by the presence of an HH in association with multisystem malformations.
   - The spectrum of features also includes pituitary hypoplasia or dysfunction, central postaxial polydactyly, dysplastic nails, bifid epiglottis, and imperforate anus.
   - In addition, cardiac anomalies, renal defects, and mild mental retardation are seen.
   - PHS is often diagnosed at birth. In familial cases it is inherited in an autosomal dominant pattern with variable expressivity.
8. *How do you classify HH? What class does this patient's hamartoma belong to?*
   - Several classification schemes have been described.
   - The most recent and most widely used is the classification of Delalande and Fohlen (▶**Fig. 71.4**).[12]
   - The present patient's lesion is classified as intrahypothalamic (3E) according to Delalande and Fohlen's classification.
9. *What treatment options can you offer to a patient with HH and intractable seizures? What are their limitations and outcomes?*
   - Microsurgical resection: seizure outcome is related to completeness of resection.[13,14]
     - Pterional and frontotemporal approach[14]
       - Advantage: shortest, most direct route to the suprasellar cistern and hamartoma
       - Disadvantage: surgical corridor may be narrowed by the internal carotid artery, optic nerve and tracts, oculomotor nerve, and pituitary stalk.
       - Outcome: 23% of patients are seizure free, 87% have significant seizure reduction (in a study of 13 patients).
     - Transcallosal interforniceal approach[15]
       - Advantage: provides a wide exposure to the third ventricle and an excellent view of the hamartoma from above; avoidance of cranial nerves and blood vessels in the suprasellar cistern and interpeduncular fossa may further reduce the risk of stroke and oculomotor nerve injury.
       - Disadvantage: risk of short-term memory deficits because of potential septal, forniceal, or mammillary body injury
       - Outcome: 52% patients are seizure free, 24% have significant improvement (in a series of 29 patients).[15]
       - Complications: thalamic infarct in 7%, increased appetite in 33% (this is permanent in 16%), and short-term memory deficits in 50%
     - Transcallosal, subchoroidal approach: alternative to the transcallosal interforniceal route; lower risk of short-term memory deficits
     - Endoscopic transventricular approach (▶**Fig. 71.5** for an endoscopic view)
       - Outcome: 31% patients were able to sustain complete resection (14 of 44 patients), of whom 90% were seizure free.[16]
       - Complications: short-term memory difficulties (three patients) and hemiparesis (one patient)[16]
   - Disconnection procedure (open or endoscopic)[17]
     - Outcome: 58% seizure free
     - Complications (in a series of 18 patients)[17]: stroke (2 patients), diabetes insipidus (2 patients), and meningitis (1 patient)
   - Stereotactic radiosurgery[18]
     - Gamma knife radiosurgery: good treatment for small- and medium-size hamartomas. The median dose recommended at the marginal isodose is 17 Gy (range is 13–26 Gy).
       - Outcome: 37% seizure free. Most had cognitive and behavioral improvement.
       - Complications: 15% transient worsening of seizures; no permanent complications mentioned
     - Linear acceleration-based radiosurgery
     - Stereotactic brachytherapy
   - Stereotactic radiofrequency ablation
     - Outcome: 25% seizure free, 25% significant improvement (12 patients)[19]
     - Complications: 8% mortality (1 patient), 16% memory deficits
   - Vagal nerve stimulation: limited role; palliative and seizure freedom is not expected[20]
   - Corpus callosotomy: limited role

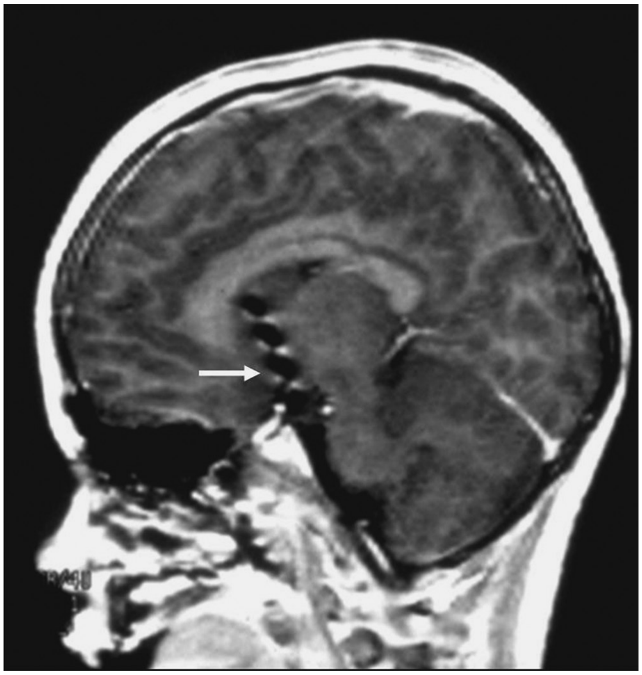
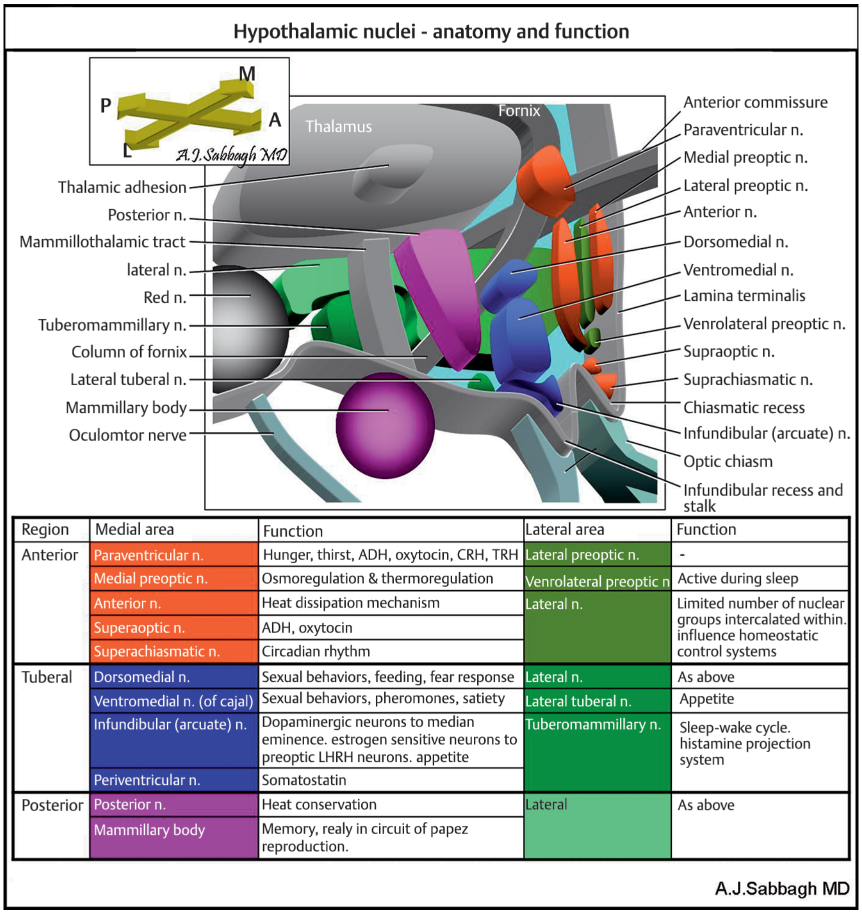


**Fig. 71.2** T1-weighted sagittal MR images of the brain showing depth electrode within the hypothalamic hamartoma.


Hypothalamic nuclei - anatomy and function
M
P
A
L
A.J.Sabbagh MD
Thalamus
Fornix
Thalamic adhesion
Posterior n.
Mammillothalamic tract
lateral n.
Red n.
Tuberomammillary n.
Column of fornix
Lateral tuberal n.
Mammillary body
Oculomtor nerve
Anterior commissure
Paraventricular n.
Medial preoptic n.
Lateral preoptic n.
Anterior n.
Dorsomedial n.
Ventromedial n.
Lamina terminalis
Venrolateral preoptic n.
Supraoptic n.
Suprachiasmatic n.
Chiasmatic recess
Infundibular (arcuate) n.
Optic chiasm
Infundibular recess and stalk
A.J.Sabbagh MD

| Region | Medial area | Function | Lateral area | Function |
|---|---|---|---|---|
| Anterior | Paraventricular n. | Hunger, thirst, ADH, oxytocin, CRH, TRH | Lateral preoptic n. | - |
| | Medial preoptic n. | Osmoregulation & thermoregulation | Venrolateral preoptic n | Active during sleep |
| | Anterior n. | Heat dissipation mechanism | Lateral n. | Limited number of nuclear groups intercalated within. influence homeostatic control systems |
| | Superaoptic n. | ADH, oxytocin | | |
| | Superachiasmatic n. | Circadian rhythm | | |
| Tuberal | Dorsomedial n. | Sexual behaviors, feeding, fear response | Lateral n. | As above |
| | Ventromedial n. (of cajal) | Sexual behaviors, pheromones, satiety | Lateral tuberal n. | Appetite |
| | Infundibular (arcuate) n. | Dopaminergic neurons to median eminence. estrogen sensitive neurons to preoptic LHRH neurons. appetite | Tuberomammillary n. | Sleep-wake cycle. histamine projection system |
| | Periventricular n. | Somatostatin | | |
| Posterior | Posterior n. | Heat conservation | Lateral | As above |
| | Mammillary body | Memory, realy in circuit of papez reproduction. | | |

**Fig. 71.3** Hypothalamic nuclei and regions. Medial and lateral areas are illustrated in shades of *red/purple* or *green*, respectively. n, nucleus; ADH, antidiuretic hormone; CRH, corticotrophin-releasing hormone; TRH, thyrotropin-releasing hormone; LHRH, luteinizing hormone release hormone; A, anterior; P, posterior; M, medial; L, lateral. (Courtesy of the Pan Arab Journal of Neurosurgery.)

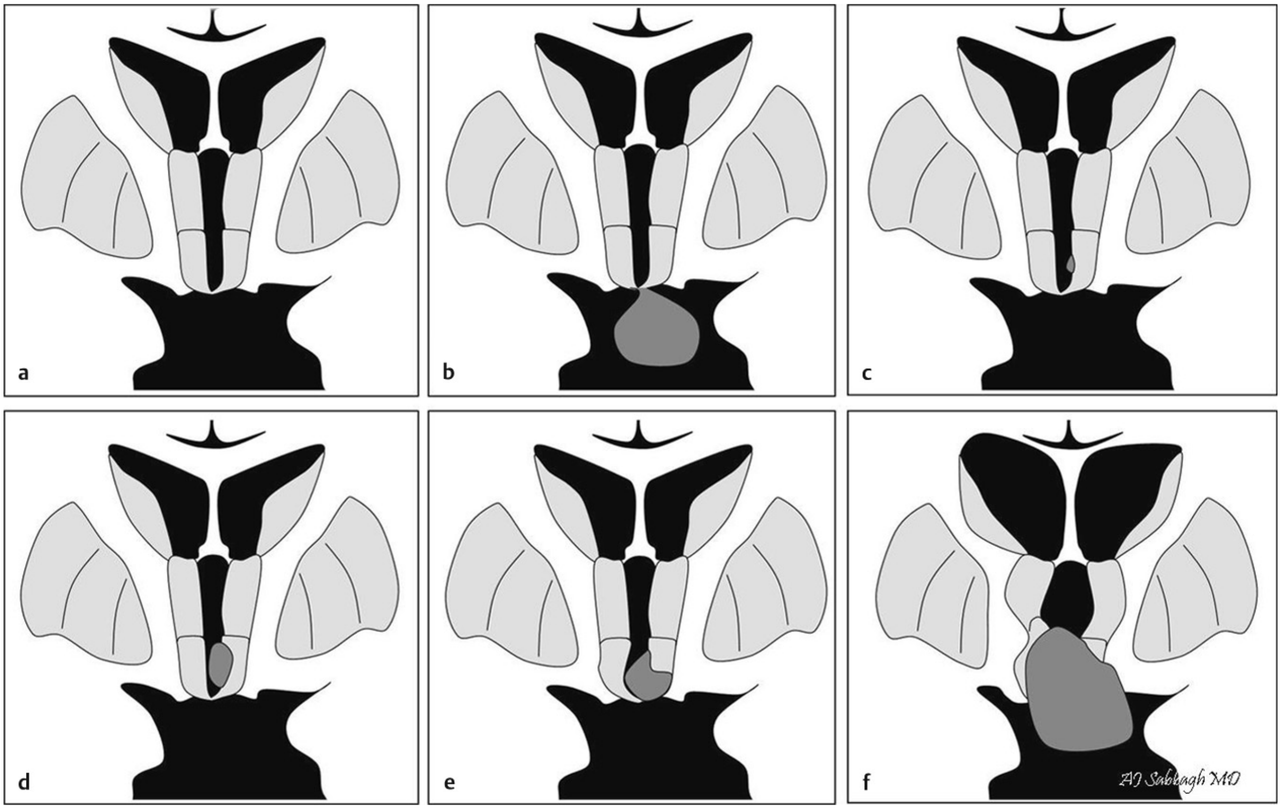


**Fig. 71.4** Classification of hypothalamic hamartomas (coronal section). (**a**) Normal; (**b**) peduncular, Delalande and Fohlen type I (horizontal insertion); (**c**) parahypothalamic, sessile Delalande and Fohlen type II (vertical insertion); (**d**) intrahypothalamic, sessile, Delalande and Fohlen type III (vertical insertion); (**e**) intrahypothalamic, Delalande and Fohlen type III (horizontal and vertical insertion); (**f**) intrahypothalamic, Delalande and Fohlen type IV (giant). (Adapted from Delalande and Fohlen 2003)[12]

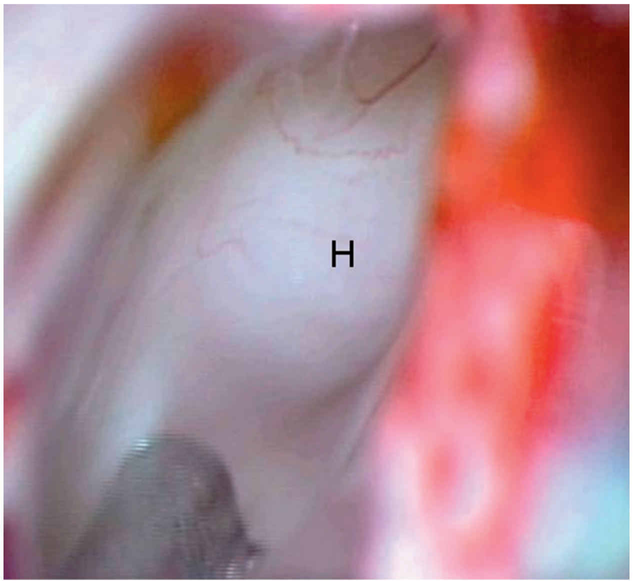


**Fig. 71.5** Intraoperative endoscopic view of a hypothalamic hamartoma (H).

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# Case 72 Posterior Fossa Juvenile Pilocytic Astrocytoma

*Asem Salma, Unwar Ul-Haq, and Essam A. Al Shail*

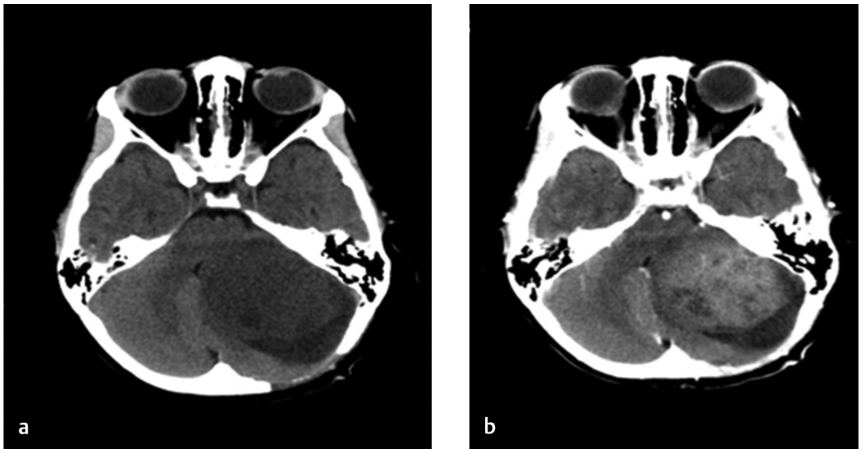

**Fig. 72.1** Brain CT, axial sections, without (**a**) and with (**b**) contrast.

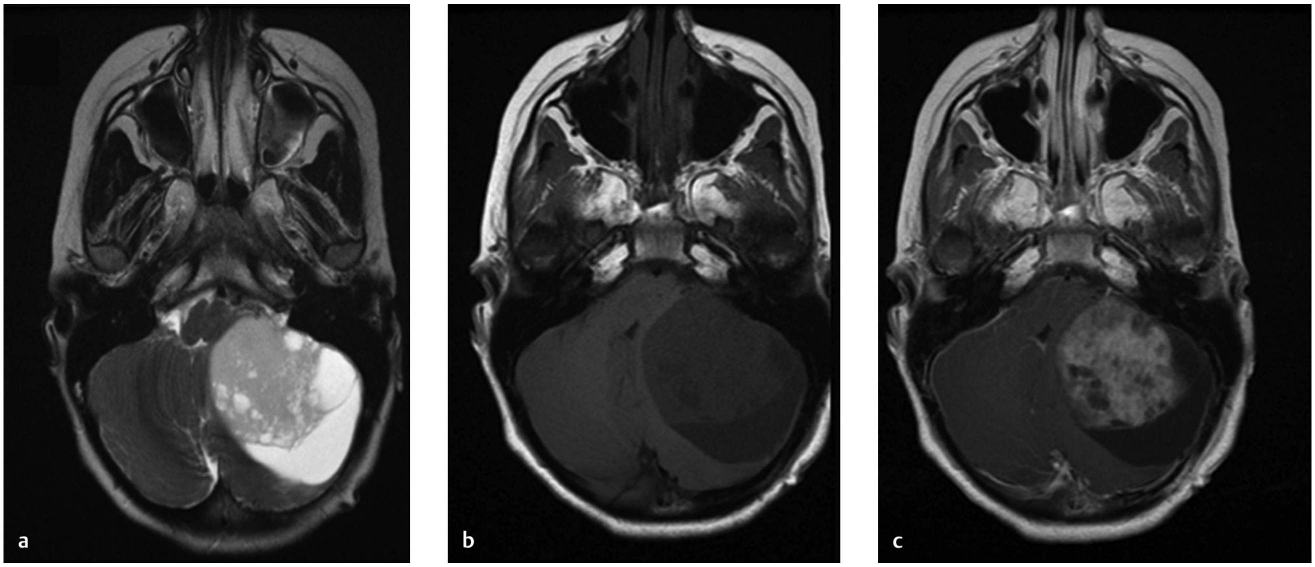

**Fig. 72.2** Brain MRI, axial sections, T2-weighted (**a**), and T1-weighted without (**b**) and with (**c**) contrast images.

## Clinical Presentation

- A 7-year-old female presents with a 6-month history of blurred vision, clumsiness, slurred speech, headache and episodes of vomiting.
- Clinical examination revealed a broad-based, ataxic gait with poor coordination.
- The patient underwent brain imaging studies that are shown in ▶**Fig. 72.1** and ▶**Fig. 72.2**.

## Questions

1. Describe the radiological characteristic of the lesion shown in ▶**Fig. 72.1** and ▶**Fig. 72.2**; what is the most likely diagnosis based on these radiological features and the clinical presentation?
2. What are the radiological patterns of pilocytic astrocytoma?
3. What is your differential diagnosis based on clinical and imaging findings of this patient?
4. What is the most worrisome radiological finding of pilocytic astrocytoma, and why?
5. Suppose this patient is 40 years old, what is the most important deferential diagnosis in this case?
6. What are the histopathological characteristics of pilocytic astrocytoma?
7. Which brain tumor syndrome can this type of brain tumor be associated with?
8. What is the most common genetic abnormality associated with pilocytic astrocytoma?
9. Name the most important considerations in the treatment plan of pilocytic astrocytoma?
10. What are the most important considerations of the postoperative care and follow-up?

## Answers

1. *Describe the radiological characteristic of the lesion shown in* ▶**Fig. 72.1** *and* ▶**Fig. 72.2***; what is the most likely diagnosis based on these radiological features and the clinical presentation?*
   - In ▶**Fig. 72.1**, brain CT scan axial sections, with and without contrast show a left large cystic posterior fossa mass. It is predominantly hypodense on the precontrast images with heterogeneous enhancement on postcontrast images.
   - ▶**Fig. 72.2** shows brain MRI, axial sections, T2-weighted and T1-weighted without and with contrast images, revealing a large cystic lesion with an enhancing mural nodule but a nonenhancing cyst wall. On T2-weighted images, the mural nodule appears hyperintense to normal brain.

   Based on these radiological features, this lesion is most likely a pilocytic astrocytoma.[1,2]
2. *What are the radiological patterns of pilocytic astrocytoma?*
   - There are three radiological patterns of pilocytic astrocytoma:[1–3]
     - Classic: a cystic lesion with an enhancing mural nodule in approximately two-third of cases. The cyst wall sometimes does enhance (46% of case) and sometimes does not (in 21% of cases).
     - False cystic astrocytoma: a cystic (central nonenhancing zone because of the necrosis) component within the tumor mass (in 16% of cases).
     - Solid: no cystic component (in 17% of cases)

   Of note, a wall thicker than 2 to 3 mm and enhancing is considered neoplastic.[3]
3. *What is your differential diagnosis based on clinical and imaging findings of this patient?*
   - The differential diagnosis should include:
     - The most common posterior fossa tumors of childhood such as:
       - Medulloblastoma
       - Ependymoma
       - Other types of cerebellar astrocytomas:
         - Pilomyxoid astrocytoma
         - Low-grade diffuse astrocytoma
         - Higher-grade astrocytoma (grade III anaplastic astrocytoma and grade IV glioblastoma multiforme).[4]
     - The relatively circumscribed tumors such as:
       - Ganglioglioma (grade 1)
       - Dysembryoplastic neuroepithelial tumor (DNET) (grade 1)
       - Rosette-forming glioneuronal tumor of the fourth ventricle (RFGNT) (grade 1)
       - Pleomorphic xanthoastrocytoma (PXA) (grade II). [3]
4. *What is the most worrisome radiological finding of pilocytic astrocytoma, and why?*
   - The most worrisome radiological finding is invasion to the brainstem, as this may prevent total resection of the tumor.
   - When dealing with pilocytic astrocytoma, the extent of the tumor resection is the most important prognostic factor:
     - A total resection is associated with overall a 5-year survival of 90%.
     - A subtotal resection is associated with 5-year survival of about 48.5%.[3]

## Answers (*continued*)

5. *Suppose this patient is 40 years old, what is the most important deferential diagnosis in this case?*
   - Hemangioblastoma, as it is also a cystic lesion with enhanced nodule; but this kind of tumor usually occur in adults with peak incidence between 40 to 60 years of age, and the wall rarely shows enhancement.[5]
6. *What are the histopathological characteristics of pilocytic astrocytoma?*
   - The histopathological picture of pilocytic astrocytoma is characterized by what is called a biphasic pattern.
   - This pattern is composed of the following:
     - Regions of dense pink–red fibrillary cells comprising spindle-shaped bipolar (piloid) astrocytes with elongated nuclei and very long and thin hair-like processes extending from both tips of tumor cells
     - Alternate with loose regions that represent microcystic and often mucinous degeneration of the tumor, as well as multipolar (and not bipolar) cells[3,5,6]
7. *Which brain tumor syndrome can this type of brain tumor be associated with?*
   - Neurofibromatosis type 1 (NF1): pilocytic astrocytoma is the most common tumor seen in patients harboring NF1, with occurrence rate up to 15 to 21% of all NF1 patients. However, in this population of patients, pilocytic astrocytoma typically involves the optic nerve or chiasm.[1,3]
8. *What is the most common genetic abnormality associated with pilocytic astrocytoma?*
   - The most frequent genetic alteration associated with pilocytic astrocytoma is a tandem duplication on the long arm of chromosome 7 (7q34 7q34) that results in *KIAA1549-BRAF* fusion.
   - This genetic change is observed in up to 70% of pilocytic astrocytoma especially in the cerebellar tumors. For this reason, the identification of the *KIAA1549-BRAF* fusion has most recently been used as a diagnostic marker for pilocytic astrocytomas.
   - In this genetic abnormality, the N-terminal end of the KIAA1549 protein substitutes the N-terminal regulatory region of BRAF, at the same time, BRAF kinase domain continues to be reserved and upregulated.[7,8]
9. *Name the most important considerations in the treatment plan of pilocytic astrocytoma?*
   - The treatment plan should always aim to achieve a complete resection of the tumor.
   - The surgery should be planned in an urgent fashion because of the risk of progressive hydrocephalus and the possible risk of acute bleeding.
   - The surgical approach varies depending on the tumor location (suboccipital craniotomy for midline lesion versus retrosigmoid craniotomy for hemispheric region posterior fossa mass.
   - Cerebrospinal fluid (CSF) aspiration maneuvers to relieve the tumor mass effects on the surrounding cerebellum should always be considered (opening the cisterna magna, a brain needle can be inserted through the dura into the cyst with subsequent aspiration of cyst fluid to achieve decompression).
   - Intraoperative ultrasound can aid in locating smaller tumors.
   - It is important to decide whether the cyst wall is neoplastic or reactive. To this end, when in doubt, multiple biopsies of the wall for frozen histology are recommended. It is preferable to leave the cyst wall in situ when it is not neoplastic.
   - Dural closure is not essential but is advised to decrease the risks for chemical meningitis. Similarly, replacing the occipital bone is advised to decrease the risk for pseudomeningocele formation and facilitate reoperation in the case of tumor recurrence. Finally, it is imperative to meticulously approximate the muscle, fascia, and superficial layers to prevent CSF leakage and pseudomeningocele formation.[3]
10. *What are the most important considerations of the postoperative care and follow-up?*
    - Postoperative MRI with and without contrast within 48 hours of the surgery should be ordered to verify the extent of resection.
    - If total resection is achieved no other treatment is suggested, and only follow-up imaging studies are advised (the recurrence rate with total resection is 0 to 6%).
    - No other adjunctive treatment is indicated with complete resection.
    - However, if only subtotal resection has been accomplished, early reoperation should be considered, should it be possible. Otherwise, the residual tumor can be followed up by regular imaging studies, as the behavior of a residual of this type of tumor is unpredictable. For example, it has been shown to stay stable or even regress in up to 45% of patients in one series at long term of follow-ups.
    - With progression the first option is to reattempting a complete surgical resection. Should this option not be possible, focal radiation such as stereotactic radiosurgery can be considered.
    - Chemotherapy is only the frontline adjuvant therapy in infants and young children (less than 5 year of age).
    - After tumor resection, the majority of patients with cerebellar astrocytoma generally demonstrate resolution of hydrocephalus. Up to 15% of patients having completed tumor resection will need to undergo a permanent CSF shunting procedure (in contrast, with medulloblastoma, where up to 50% of patients will need shunt placement).[3]

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# Case 73 Vein of Galen Malformation

*Matthew Pierson, Randall C. Edgell, and Samer K. Elbabaa*

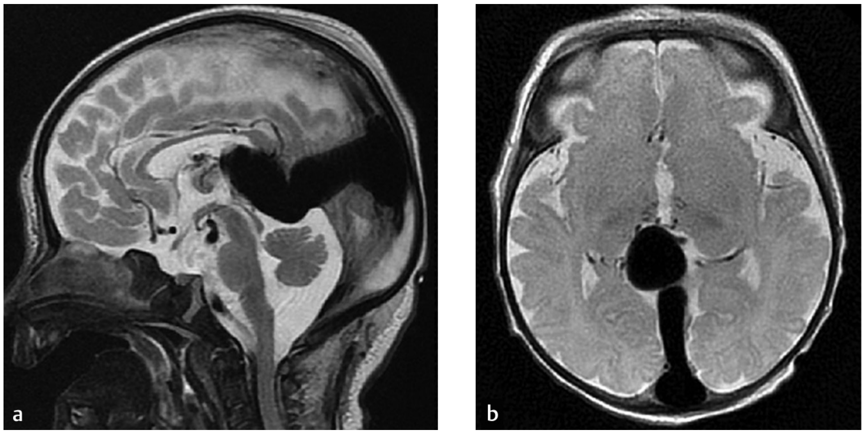

**Fig. 73.1** Depicted are T2-weighted sagittal (**a**) and axial (**b**) MR images of the brain.

## Clinical Presentation

- A 3-day-old male infant is delivered via Cesarean section with an unremarkable prenatal history. The infant was intubated for respiratory distress and a head ultrasound performed revealed a vascular malformation.
- Physical examination showed an awake infant with a good cry, pupils are equal and reactive, fontanelle is soft and flat, and there is no splaying of the skull sutures. A faint bruit is heard on auscultation of the anterior fontanelle. The head circumference measures 37.8 cm.
- Head ultrasound demonstrates a cystic lesion near the quadrigeminal cistern which exhibits high flow on color Doppler. The ventricles are of normal size and no hemorrhage is observed.
- MRI is obtained and shown below (▶**Fig. 73.1**).

## Questions

1. Interpret the MRI findings.
2. What further diagnostic studies and work-up would you like to obtain?
3. What is the most likely diagnosis?
4. Describe the incidence of this lesion.
5. What conditions do you need to evaluate when considering treatment options? Discuss the treatment options.
6. Interpret the initial cerebral angiogram provided (▶**Fig. 73.2**).
7. You decide to proceed with endovascular embolization of the lesion. What is the indication for treatment? What is the goal of the treatment? Does the age of presentation affect your treatment plan?
8. Interpret the post embolization angiogram images (▶**Fig. 73.3**).
9. How will you follow the patient after treatment?
10. Provide a radiographic classification of this pathology.

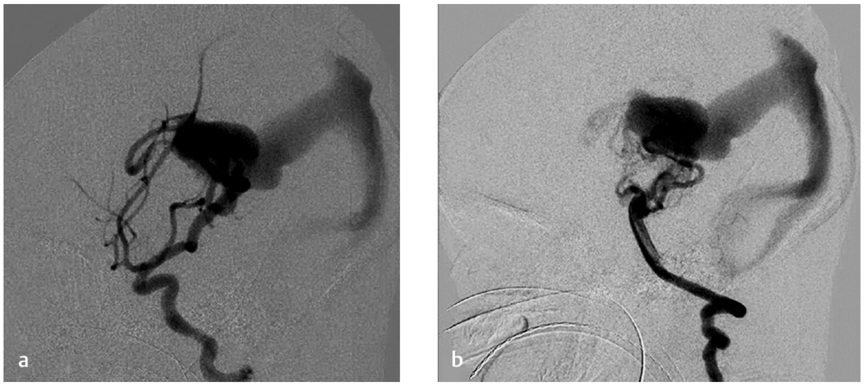

**Fig. 73.2** Lateral images of a cerebral angiogram during the (**a**) carotid injection and (**b**) vertebral artery injection.

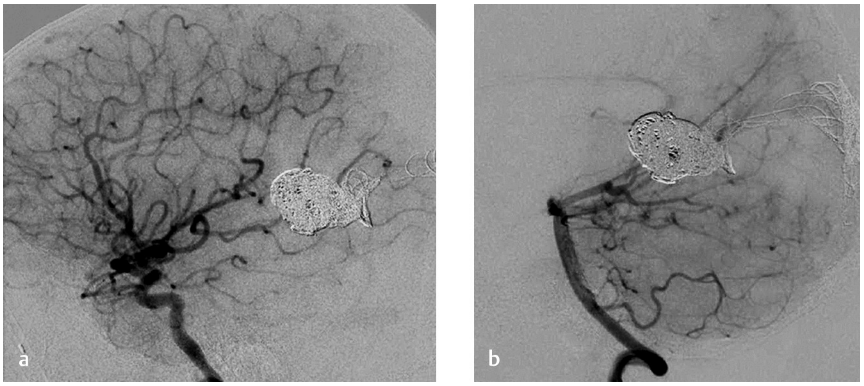

**Fig. 73.3** Lateral images of a cerebral angiogram during the (**a**) carotid injection and (**b**) vertebral artery injection after endovascular embolization.

## Answers

1. *Interpret the MRI findings.*
   - There is dilation of the vein of Galen, falcine sinus, and confluence of sinuses without ventriculomegaly.
2. *What further diagnostic studies and work-up would you like to obtain?*
   - Pediatric cardiology consultation and echocardiogram to evaluate for high-output congestive heart failure
   - Close monitoring of head circumference
   - Four-vessel angiogram and concomitant endovascular treatment if there is clinical heart failure, hydrocephalus, or change in neurologic exam
3. *What is the most likely diagnosis?*
   - Vein of Galen malformation (VOGM)
   - This is a dilation of the median vein of the prosencephalon, which is a precursor of the vein of Galen, caused by an arteriovenous (AV) shunt from the choroidal arteries of the anterior and posterior circulation.[1]
4. *Describe the incidence of this lesion.*
   - The true incidence is unknown, but VOGMs are rare.
   - The reported incidence is less than 1% of cerebral vascular malformations and 30% of pediatric vascular malformations.[2]
   - The first description of a VOGM was probably by Steinhill in the German pathology literature in 1895.[3]
5. *What conditions do you need to evaluate when considering treatment options? Discuss the treatment options.*
   - Most conditions encountered in VOGM malformations are related to the degree of AV shunting.
   - High-output congestive heart failure (if present): Management by a pediatric cardiologist is critical to evaluate the severity of heart failure and need for medical treatment (digoxin, diuretics, ventilator support). Emergent endovascular therapy may be needed to reduce the AV shunt, which can improve cardiac function.
   - Ventriculomegaly: This results from increased dural sinus pressure secondary to AV shunting and occasionally due to compression of the cerebral aqueduct.
     - The increase in dural sinus pressure creates a resistance for cerebrospinal fluid (CSF) to enter the sinus, which may lead to hydrocephalus and intracranial hypertension.
     - Close monitoring of head circumference, apneas, bradycardias, and the status of the fontanelle is important for possible signs of hydrocephalus.
     - Generally, CSF shunting without treatment of the VOGM is not recommended because of high risk of hemorrhage and seizures.[4]
     - If the increase in head circumference seems too rapid or the clinical follow-up period demonstrates a significant developmental delay, urgent

## Answers (*continued*)

embolization should be performed and ventricular shunting should be avoided.[5]

- Seizures (if present): There may be a need for antiepileptic therapy in the neonatal period.
- Treatment options are discussed below.
  - Children with VOGMs have very poor prognosis if left untreated. An extensive clinical and anatomic evaluation is needed to determine the optimal timing of treatment. A short period of observation may improve the outcome if clinically tolerated.
  - Surgical clipping: Open microsurgical clipping was the best available treatment option before the era of endovascular therapy.
    - Different techniques have been described.[3,6,7]
    - Yasargil et al emphasized the utility of a posterior interhemispheric approach in a sitting position.[6]
    - The deep central fistulous communications that are characteristic for VOGM make them less amenable to excision than endovascular treatment.[8]
  - Endovascular embolization: Advances in endovascular techniques and imaging have helped us understand the anatomy and pathophysiology of VOGM. They have also provided new treatment options with improved outcomes.
  - Circillo et al in 1990 retrospectively compared the outcomes of both surgical and endovascular techniques in neonates and concluded that endovascular options are far safer.[9]
  - Endovascular treatment has become the cornerstone for good outcomes in these challenging lesions.
  - Lasjaunias et al reported a large series of 317 patients. Of these, 233 patients were treated with endovascular therapy from 1981 to 2002. The treatment method of choice was a transfemoral arterial approach to deliver glue at the fistulous site. In their series, mortality rate after embolization was 10%. Of the surviving patients, moderate to severe developmental delay was seen in 26% of patients; however, 74% were neurologically normal on follow-up.[5]
  - Gamma knife radiosurgery: This treatment modality is being increasingly reported as providing definitive therapy in select patients.
  - Payne et al reports a near 50% cure rate in a series of nine patients with no morbidity or mortality related to the radiosurgery.[10]
  - Radiosurgery may be used in conjunction with other therapies or potentially as a standalone modality in clinically stable patients.[10,11]

6. *Interpret the initial cerebral angiogram provided (▶* **Fig. 73.2***).*
   - The images show aneurismal dilation of the median vein of the prosencephalon on both carotid and vertebral artery injections.
   - The main feeders are choroidal vessels from the anterior and posterior circulation.
7. *You decide to proceed with endovascular embolization of the lesion. What is the indication for treatment? What is the goal of the treatment? Does the age of presentation affect your treatment plan?*
   - The indications for treatment include high-output congestive heart failure, hydrocephalus, or neurologic symptoms.
   - The primary goal of treatment is to control the AV shunt. A staged embolization is generally recommended for neonates.
     - This provides preservation of the hydrovenous equilibrium.
     - It limits high renal exposure to contrast loads as well as radiation time.
     - Severe encephalomalacia and multiorgan system dysfunction are associated with a poor outcome despite a technically successful embolization in some patients when a single stage embolization is performed.
     - In infants, normal brain development should be preserved by endovascular exclusion of the fistula if technically feasible.[2,5]
     - In 2006, Lasjaunias et al summarized 20 years of experience in the treatment of VOGM at Le Kremlin Bicetre in Paris, France. They emphasized the importance of a thorough analysis of neonatal patients with VOGM to best predict the degree of cerebral tissue impairment not evident on imaging.[5]
       - The VOGM neonatal score was developed by their group, which included cardiac, cerebral, respiratory, hepatic, and renal functions.
       - Their decisions follow a strict protocol based on the neonatal score derived from the above information.[5]
8. *Interpret the post embolization angiogram images (▶* **Fig. 73.3***).*
   - The fistula has been successfully obliterated via transvenous coil embolization.
9. *How will you follow the patient after treatment?*
   - Continue follow-up with pediatric cardiology with echocardiogram evaluation for residual heart failure.
   - Head circumference
   - Monitor for radiographic evidence of hydrocephalus via head ultrasound, CT, or MRI.

## Answers (*continued*)

- Follow-up cerebral angiogram at 6 months post treatment. If contrast hyperemia is present at 6 months, even without AV shunting, follow-up angiograms are recommended at 1 and 2 years later.[5]

10. *Provide a radiographic classification of this pathology*.
    - The most widely referenced angiographic classifications are those of Lasjaunias et al and Yasargil et al.[6,12]
    - Lasjaunias et al classification includes true and secondary VOGM.
      - The true VOGM occurs because of a dysembryogenic event involving the median vein of the prosencephalon.
        - As a result, a fistulous connection develops between the choroidal arteries and the veins in the wall of the dilated vein of prosencephalon.
        - The deep venous system has a separate drainage pattern without communicating with the fistula.
        - Further subcategories of the true VOGM include the mural type where the fistula is in the wall of the median vein of the prosencephalon, or the choroidal type with drainage into tributary veins of the median vein of the prosencephalon.
      - The secondary types of VOGMs have adjacent AV malformations that drain selectively into the great vein of Galen.
        - These lesions are characteristically supplied by branches of the middle cerebral artery (thalamoperforators, lenticulostriate, or transsylvian branches), whereas the choroidal and mural true VOGMs are supplied by choroidal and pericallosal vessels.[13]
    - Yasargil et al. defined four types of VOGM lesions:
      - Type 1 is a simple fistula involving branches from the pericallosal or posterior cerebral arteries.
      - Type 2 involves more feeding branches from the middle cerebral artery branches.
      - Type 3 involves high-flow lesions with large numbers of fistulous connections from a wide range of feeding vessels.
      - Type 4 is a midline AV malformation with drainage into the vein of Galen (analogous to the Lasjaunias secondary VOGM).[12]
    - Of note, a scoring system has also been proposed by Mortazavi et al based on age and presence of heart failure rather than radiographic anatomical classification.[12]

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# Case 74 Pediatric Head Trauma

*Jeffrey Atkinson, José Luis Montes, and Abdulrahman J. Sabbagh*

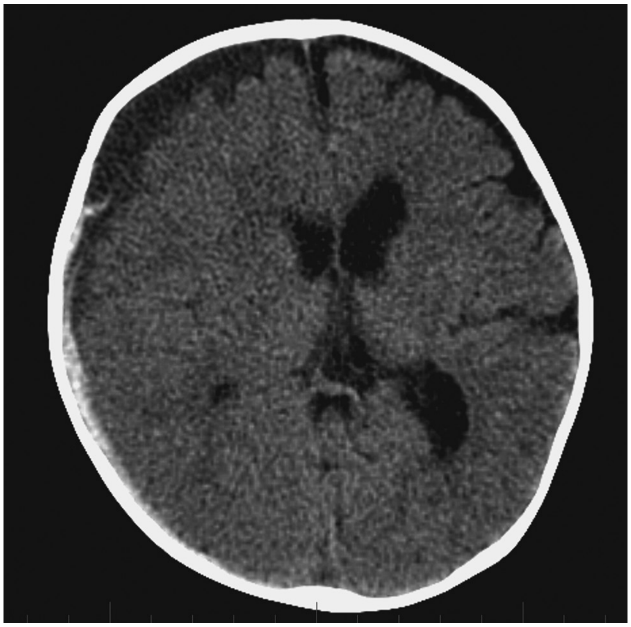

**Fig. 74.1** CT scan of the brain revealing extra-axial fluid collection along the right convexity.

## Clinical Presentation

- A 7-month-old child was found at home with decreased level of consciousness and no clear history of antecedent event.
- He was transported to the hospital where he had a focal seizure.
- Initial examination reveals a Glasgow Coma Score (GCS) of 5, with questionable motor asymmetry and a tense and bulging fontanel.
- He is intubated urgently and sent for a CT scan. A representative image appears in ▶**Fig. 74.1**.

## Questions

1. What are the findings on the CT scan?
2. What is the differential diagnosis?
3. What other points on history and physical examination might be important?
4. What other investigations and consultations are relevant?
5. What is the acute management of the patient?

   The patient was taken urgently to the operating room for decompression and evacuation of the subdural hematoma (SDH). He recovered well from the injury.

6. What are the principles of intracranial pressure (ICP) management in a child?
7. What is the epidemiology of inflicted trauma?
8. Is there a constellation of clinical and neurologic findings that are often associated with nonaccidental trauma?
9. Are retinal hemorrhages pathognomic of nonaccidental trauma?
10. What is the differential diagnosis with the association of retinal hemorrhage and bilateral SDHs?
11. What are the most common findings associated with nontraumatic head injury on CT scan?
12. What are the mechanisms of head injury involved in infants and young children?
13. What is the prognosis in a child with an abusive head injury?
14. What are the medicolegal implications of inflicted trauma?

## Answers

1. *What are the findings on the CT scan?*
    - The CT scan of the head demonstrates subdural collections over the right hemisphere, which are both acute and chronic.
    - There is associated mass effect on the adjacent hemisphere with compression of the ipsilateral ventricle, loss of sulcations, and midline shift.
2. *What is the differential diagnosis?*
    - This SDH is most probably traumatic.
    - The acute and chronic components may indicate hemorrhages of different ages or they may indicate an acute SDH on top of an enlarged subarachnoid space such as in the syndrome of benign extra-axial fluid collection of infancy.
    - Nontraumatic causes of SDH are very rare in infants but might include vascular malformation (of which there is no evidence on these images), coagulation disorder, or inborn error of metabolism.
3. *What other points on history and physical examination might be important?*
    - Important information obtained on history includes:
        - History of trivial blunt trauma often from falling from beds or low chairs or no history of trauma at all.
        - Decreased level of consciousness from lethargy to coma.
        - Irritability, vomiting, seizures, apnea, hypoxia
    - Important findings on inspection include:
        - An enlarged head
        - Unusual or patterned bruising
        - Venous engorgement
    - Important neurologic findings on examination include:
        - Abnormal movements
        - Seizure activity
        - Tense fontanelle
        - Retinal hemorrhages
        - Lateralizing findings
    - Given the likely traumatic nature of this injury and the suspicion of hemorrhages of different ages, a detailed history must be taken. We need to better establish the exact nature of any traumatic injury the child might have received and establish a time line for caregivers (i.e., exactly who was caring for the child and when).[1]
    - In addition, a social history from the family would be relevant, as well as any previous history of hospital visits for suspicious injuries.[1–3]
4. *What other investigations and consultations are relevant?*
    - Consultations should be obtained from the local child abuse investigative team, which might include pediatrics and social work.[1]
    - Ophthalmology should be consulted to examine the fundi properly to look for retinal hemorrhages that might further suggest abusive injury.[4]
    - Skeletal survey or nuclear medicine bone scan should be done to look for fractures.[4]
    - MRI of the brain might be considered to better evaluate for cortical or parenchymal injury to the brain and perhaps to better define the ages of the subdural blood.
    - A standard investigation of abnormal coagulation would also be of benefit.[1]
5. *What is the acute management of the patient?*
    - Acute management of this patient involves airway protection as already established by intubation.
    - ICP control medically can be attempted by mannitol, controlled ventilation, sedation of the child, which would imply the insertion of an ICP monitor.[5]
    - Seizure control should be obtained by an intravenous load of the antiepileptic of choice, usually phenytoin or phenobarbital.
    - In this case with the mass effect, the clinical evidence of high ICP and the clinical state of the patient, the argument for evacuation of this clot can be made. This is probably best done by craniotomy, though burr hole or transfontanel evacuation of the chronic component could be considered.
    - An ICP monitor should be inserted into the patient at the time of surgery for postoperative management.
6. *What are the principles of intracranial pressure (ICP) management in a child?*
    - The principles of ICP management in a child are very much the same as those in an adult.
    - A child such as this patient with a GCS < 8 and a positive CT scan should be monitored invasively.[5]
    - Even in an infant such as the one in this case, physical examination of the fontanelle is not necessarily adequate for monitoring.
    - With the clot evacuated and the monitor in place, controlled ventilation, cerebrospinal fluid (CSF) drainage if possible, sedation, and osmotic agents can be used.[6]
    - In children, there is also evidence of the benefit of continuous infusions of hypertonic saline in addition to mannitol.[7]
7. *What is the epidemiology of inflicted trauma?*
    - Population-based studies have indicated an annual incidence of 24.6 per 100,000 children younger than 1 year of age.[8]
    - The risk of suffering an inflicted head injury by 1 year of age has been established at 1 in 4,065.[8]
    - In most studies, boys are slightly more at risk than girls.[9]
    - Some of the risk factors include young parents, low socioeconomic status, single parents, prematurity of the infant, history of abuse by the caretaker, and a history of psychiatric or substance abuse.
    - The most common perpetrators are fathers and boyfriends.
    - Female babysitters account for 18% of perpetrators.

## Answers (*continued*)

8. *Is there a constellation of clinical and neurologic findings that are often associated with nonaccidental trauma?*
   - Yes, the association consists of:
     - Retinal hemorrhage and subarachnoid hemorrhage or SDHs[10]
     - Bilateral chronic SDHs
     - On CT scan, the association between subarachnoid hemorrhage, SDH, and interhemispheric blood is highly suspicious of this entity.
9. *Are retinal hemorrhages pathognomic of nonaccidental trauma?*
   - No, they can be associated with normal vaginal delivery.
   - Other factors associated with retinal hemorrhages include accidental trauma, coagulopathy, hypertension, subarachnoid hemorrhage, subdural hemorrhage, papilledema, arterial hypertension, and resuscitation.[11]
10. *What is the differential diagnosis with the association of retinal hemorrhage and bilateral SDHs?*
    - Accidental trauma
    - Osteogenesis imperfecta
    - Blood coagulation dyscrasias
    - Metabolic disorders such as glutaric acidemia type I[11]
11. *What are the most common findings associated with nontraumatic head injury on CT scan?*
    - Acute SDH[9]
    - Interhemispheric hemorrhage, particularly posterior or layering the tentorium
    - Parenchymal hypodensities sometimes presenting as a black brain, but most common as hemispheric or patchy hypodensities
12. *What are the mechanisms of head injury involved in infants and young children?*
    - The development of acute SDH with parenchymal contusions and significant symptoms is most likely a combination of tangential acceleration, usually associated with shaking episodes and impact manipulations that are caused by hitting the head of the child against a blunt surface or throwing him against one.
    - The forces associated to impact manipulation are in the order of 20 to 30 times greater than the forces generated by shaking alone; the time lapse is significantly shorter.
    - The forces necessary to develop acute SDHs, brain contusion, and diffuse axonal injury are most likely related to impact manipulation or a combination of both.
    - Low-height free-fall forces are enough to cause skull fractures or nonsymptomatic subdural hemorrhages, but not acute subdural hemorrhage accompanied by significant acute neurologic deficit as observed in most nonaccidental trauma.
    - Occasionally, low-height free falls may cause epidural hematomas that are highly symptomatic, but this represents the exception.[9,12,13]
13. *What is the prognosis in a child with an abusive head injury?*
    - In general prognosis with abusive head injury is quite poor, probably due to extensive cortical and parenchymal injury of the associated brain and even cervical medullary junction.[9,14]
    - There is an overall 15 to 38% mortality rate. Thirty to 50% of survivors will have cognitive or other neurological deficits, and ~30% will have no significant sequelae.[9]
    - However, in this case there appears to be very little cortical damage on the CT scan. There needs to be MRI and clinical confirmation. There might be reason to be optimistic if the clinical evolution after surgery is positive.
14. *What are the medicolegal implications of inflicted trauma?*
    - The neurosurgeon is often called to establish whether there is evidence of nonaccidental trauma.
    - The neurosurgeon should be in close contact with the child protection team and ensure that all tests and studies have been done to document and support the diagnosis.
    - The neurosurgeon should be forthcoming with his or her opinion after careful consideration. Unfortunately, in medicine, answers are sometimes not clear-cut and proof of child abuse may be difficult to establish.

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# Case 75 Pediatric Intracranial Epidural Abscess

*Exequiel P. Verdier and Samer K. Elbabaa*

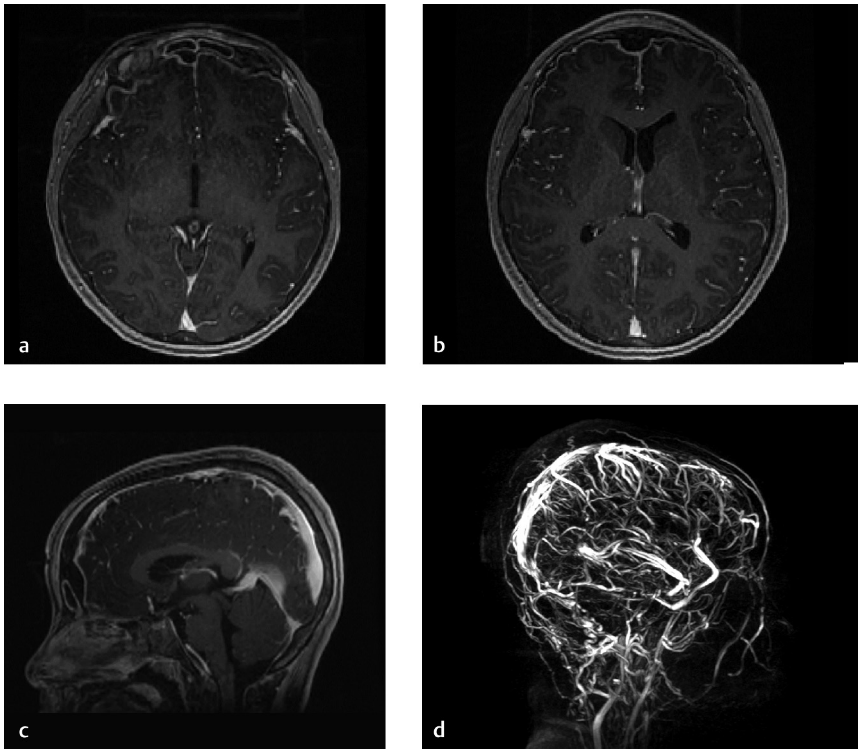

**Fig. 75.1** Axial T1-weighted images with contrast (**a**) and (**b**), midsagittal T1-weighted image (**c**), and MR venogram (**d**) of the patient herein.

## Clinical Presentation

- An 8-year-old girl presents with a discreet swelling of the forehead, fever, lethargy, and headaches.
- She has a registered history of several frontal sinusitis.
- Laboratory studies show: white blood cells = 14,500/mm$^3$, C-reactive protein (CPR) elevated, blood cultures negative.
- MRI with contrast and MR venography (MRV) is performed and shown in ▶**Fig. 75.1**.

## Questions

1. Describe the MRI pictures.
2. What is the most likely diagnosis for this case? Provide a differential diagnosis.
3. Which other studies can you do to confirm the diagnosis?
4. Provide epidemiology and microbiology of epidural abscess.
5. Describe the pathophysiology. What are the routes of spread and risk factors?
6. Detail the clinical presentation and symptoms.
7. Specify initial management and treatment options.
8. Outline the complications.
9. Describe the outcomes.

## Answers

1. *Describe the MRI pictures.*
   - This is a T1-weighted MRI with gadolinium contrast that shows a biconvex low-intensity lesion in the right frontal epidural space with a patchy and leptomeningeal enhancement, associated with peripheral scalp subgaleal contrast enhancement (called Pott's puffy tumor). There is also a mucosal swelling of the frontal sinus.
   - On the MRV, you can observe superior sagittal sinus thrombosis (as a complication) at the level of the anterior third of the sinus.
   - The MRI offers much more anatomical and physiologic information than CT scans. An advantage of CT is precise demonstration of the osseous anatomy of the frontal sinus which can be helpful for endoscopic sinus surgery planning.
2. *What is the most likely diagnosis for this case? Provide a differential diagnosis.*
   - Differential diagnosis includes:
     - Brain neoplasm
     - Granuloma
     - Cerebral infarct
     - Intraparenchymal brain abscess
     - Subdural and epidural abscess
     - Chronic epidural hematoma
     - Sinusitis
   - The most likely diagnosis is epidural abscess involving the frontal sinus. An epidural abscess is a suppurative infection of the epidural space (collection of pus between the dura mater and the overlying bone).
3. *Which other studies can you do to confirm the diagnosis?*
   - If intracranial infection is suspected, contrast must be administrated in every case. CT has lower sensitivity than MRI, therefore MRI is the preferred study for detection of small collections along the epidural space.
   - Diffusion-weighted imaging (DWI) will show high-signal-intensity structures, and low intensity on apparent diffusion coefficient (ADC) maps. These changes in the frontal bone signal on DWI are suggestive of osteomyelitis.
   - MR spectroscopy will show elevation of lactate, acetate, succinate, and amino acid concentration; all these are bacterial metabolites.
4. *Provide epidemiology and microbiology of epidural abscess.*
   - The incidence of intracranial epidural abscess is unknown and uncommon; it accounts for 2 to 5% of all the intracranial suppuration. In fact, 90% of epidural abscesses occurs in the spine. It is a condition more frequently seen in mid-adolescence associated to sinus and ear infections, and has a male predominance.
   - The frontal sinus is the last paranasal sinus to develop and it becomes visible on X-ray at age 6, enlargement continues throughout childhood. The sinus becomes fully developed in adolescence.
   - Staphylococcus *aureus* and *Staphylococcus epidermidis* organisms are the most commonly isolated culprits; however, enteric gram-negative bacilli (e.g., *Escherichia coli*), *Pseudomonas* species, Bacteroides species, and other anaerobes can also be involved.
   - Aerobic and microaerophilic *Streptococci* are usually responsible for an infection that has spread from the paranasal sinuses. Gram-negative organisms (*Pseudomonas*) are prevalent in cases of middle ear suppuration associated with cholesteatoma.
5. *Describe the pathophysiology. What are the routes of spread and risk factors?*
   - Intracranial epidural abscess may result from spread of infections from the paranasal sinuses, middle ear, or orbit; usually the frontal and ethmoid sinusitis are involved in frontal abscesses (as in this case). The infection can lead to a subdural empyema by crossing emissary veins, but it may rarely occur as a result of hematogenous seeding.
   - Male sex and teenagers seems to have a vulnerability for intracranial complications of paranasal sinusitis. Chronic sinusitis and repeated middle ear disease are also risk factors. A large cohort study in the United States identified a strong association of asthma with sinogenic intracranial suppuration.
   - Other predisposing conditions for intracranial suppuration and cerebral abscess include the following:
     - Immunosuppression
     - Diabetes
     - Steroid use
     - Alcoholism
     - Renal failure
     - Intravenous drug abuse
     - Meningitis
     - Sinusitis
     - Mastoiditis
     - Penetrating trauma
     - Craniotomy
     - Dental abscess surgery
     - Congenital heart disease (right to left shunting, patent foramen ovale)
     - Pulmonary infections (lung arteriovenous fistula, Osler–Weber–Rendu syndrome)
   - Routes of spread include: (See ▶**Table 75.1**)
     - Contiguous infection (25–50%): otogenic and paranasal infections, cranial osteomyelitis
     - Hematogenous spread (15–30%): endocarditis, intra-abdominal infection, pulmonary origin, urinary tract infection

**Table 75.1** Routes of spread and antibiotic options for epidural abscess

| Routes of Spread | Microbial Agent | Antibiotics Options |
|---|---|---|
| Contiguous infection (25–50%) | | |
| Otogenic | Streptococcus species<br>Bacteroides species<br>Enterobacteriaceae<br>Pseudomonas species | Penicillin G<br>Metronidazole<br>Third-generation cephalosporins |
| Paranasal | Streptococcus species<br>Peptococcus species<br>Fusobacterium species<br>Bacteroides species<br>Propionibacterium species | |
| Cranial osteomyelitis | Staphylococcus species (aureus, epidermidis)<br>Escherichia coli (neonates) | |
| Hematogenous spread (15–30%) | | |
| Endocarditis | Viridans Streptococci<br>Staphylococcus aureus | Oxacillin<br>Metronidazole<br>Third-generations cephalosporins |
| Intra-abdominal infection | *Klebsiella pneumoniae*<br>E. Coli<br>Enterobacteriaceae<br>Streptococcus species<br>Anaerobes | Penicillin G<br>Metronidazole<br>Third-generation cephalosporins |
| Pulmonary origin | Streptococcus species<br>Fusobacterium species<br>Corynebacterium species<br>Peptococcus species | |
| Urinary tract infection | Enterobacteriaceae<br>Pseudomonas species | |
| Postneurosurgical and posttraumatic (8–19%) | | |
| Penetrating trauma | *Staphylococcus aureus*<br>Clostridium species<br>Enterobacteriaceae<br>Bacteroides species<br>Fusobacterium species | Oxacillin<br>Metronidazole<br>Third-generation cephalosporins |
| Postoperative | *S. aureus*<br>*S. epidermidis*<br>Enterobacteriaceae<br>Pseudomonas species | Vancomycin<br>Third-generation cephalosporins |

## Answers (*continued*)

  - Postneurosurgical and Posttraumatic (8–20%): direct contamination from penetrating trauma, contamination after cranial surgery.

6. *Detail the clinical presentation and symptoms.*
   - The initial symptoms are those related to the infectious process. It has an insidious onset, usually weeks to months. Headaches, fever, lethargy, sinus pain or tenderness, and sometimes forehead swelling (Pott's puffy tumor) are the basic clinical presentation symptoms and signs.
   - Fever may be persistent and headaches are at first located on the site of infection. If no treatment is initiated, the epidural suppuration enlarges gradually causing symptoms and signs of increased intracranial hypertension: vomiting, papilledema, focal neurological deficit and if the infection reaches the subdural space, seizures and neck stiffness can manifest.
   - Sixth nerve palsy can be seen as a result of elevated intracranial pressure (ICP), but also as a complication of middle ear infection (Gradenigo's syndrome: retro-orbital pain and abducens nerve palsy).
7. *Specify initial management and treatment options.*
   - It is essential to perform a clinical evaluation, chest X-rays, cardiac echography (to rule out congenital heart disease), blood count and culture before starting any treatment.
   - Sedimentation rate and CRP should also be performed.
   - Lumbar puncture is contraindicated and typically shows a low rate of bacteria growing in the sample.

## Answers (*continued*)

- Treatment options include a combination of intravenous antibiotics, sinus surgery, and neurosurgical drainage.
  - The primary treatment of intracranial suppurations is parenteral antimicrobial therapy after identifying the causative infectious agent and performing antibiotic sensitivity testing. The administrations of antibiotics before a specimen can be obtained may compromise bacterial growth in cultures and make the correct choice of antibiotics more difficult.
  - However, if there is no possibility to isolate the organism, empiric antimicrobial therapy should be initiated based on the anatomic location of the abscess (see ▶**Table 75.1**).
  - Commonly three antibiotics must be used for empirical treatment to cover common gram-positive organisms (*Staphylococci* and *Streptococci*), gram-negative organisms, and anaerobes for up to 6 to 8 weeks. This initial therapy should begin with board-spectrum antibiotics such as a third-generation cephalosporin, metronidazole, and vancomycin; but when antibiotic sensitivity reports become available, specific bactericidal agents to the organism(s) cultured should be administered.
- There is no controlled randomized clinical study that evaluates use of corticosteroids in this setting. Some studies have come to the conclusion that steroids are recommended for reducing ICP after initiation of antimicrobial therapy. One should keep in mind that steroids can also reduce the effectiveness of the immune system in fighting infections and therefore could potentially negatively impact the treatment.
- Anticonvulsant therapy is not generally recommended for epidural abscess. It is typically indicated, other than for seizure treatment, when the abscess is complicated with subdural empyema or brain abscess.
- Indications for neurosurgical treatment depend on the clinical presentation of each patient, but in every case, it is necessary to obtain pus for microbiological diagnosis.
  - The guidelines for surgical management are:
    - ○ To reduce ICP
    - ○ To obtain pus for microbiological confirmation
    - ○ To enhance antibiotic therapy
    - ○ To avoid spread of the infection to the subdural space
  - Different techniques have been described to evacuate an intracranial epidural abscess. These range from simple burr hole drainage, stereotactic needle aspiration, to performing a large craniotomy.
  - Simple burr hole drainage presents a higher risk for recurrent abscess when compared to craniotomy. Craniotomy flaps techniques are preferred when patients present with increased ICP or sings of frontal bone osteomyelitis.
  - An alternative is to perform a minimally invasive endoscopic transnasal approach to the frontal sinus with the advantage to treat the sinusitis and intracranial abscess in the same procedure with better cosmetic results and lower postoperative morbidity than traditional approaches.

8. *Outline the complications.*
   - Increased ICP
   - Spread of infection
   - Dural sinus thrombosis
   - Subdural empyema
   - Brain edema, ischemia, and/or intraparenchymal abscess
   - Osteomyelitis
   - Hydrocephalus and ventriculitis
   - Seizures
   - Recurrence (3–25%)
9. *Describe the outcomes.*
   - With the combinations of new MRI techniques and advanced antibiotics, mortality rates are reported in between 10% and nil.
   - Signs of good outcomes are young age, no neurological deficits at presentation, no comorbid factor, and early and accurate diagnosis.
   - Poor prognosis is associated with signs of high ICP at presentations, septic shock complication, and late diagnosis.
   - Complete resolution could take up to 12 weeks.
   - Follow-up is needed with neuro-imaging every 3 month to discharge recurrence.

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# Case 76 Spontaneous Cerebrospinal Fistula

*Jeffrey Atkinson, José Luis Montes, and Abdulrahman J. Sabbagh*

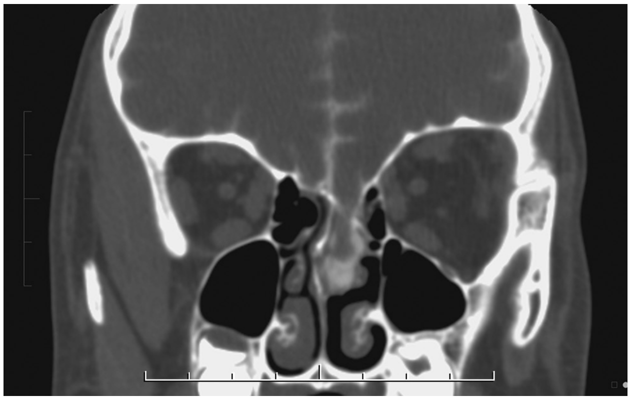

**Fig. 76.1** CT coronal scan through the anterior fossa and ethmoids, with infusion of metrizamide intrathecal contrast.

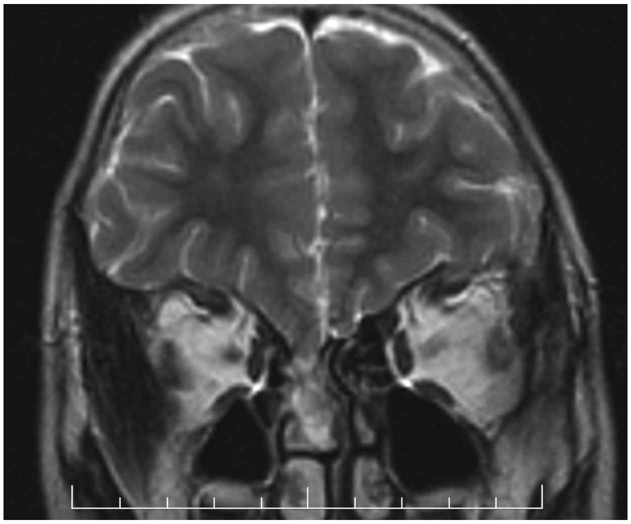

**Fig. 76.2** T2-weighted coronal MR image through the anterior cranial fossa.

## Clinical Presentation

- A 5-year-old boy presents with his third episode of bacterial meningitis.
- The child has a history of prematurity with a mild hemiparesis and mild developmental delay.
- There is no history of trauma.
- Physical examination reveals no skin defects or cutaneous markers.
- On detailed questioning, the child does recall that his nose is frequently "runny," and with prolonged forward positioning small amounts of clear fluid can be found dripping from the nose.

## Questions

1. What are the potential causes of spontaneous cerebrospinal fluid (CSF) fistula?
2. Where are the potential sites of CSF leak?
3. What other clinical syndromes might result from CSF fistula?
4. What would be the diagnostic tests indicated in this patient?

   The patient went on to have MRI and CT scans following intrathecal infusion of metrizamide that are seen in ▶**Fig. 76.1** and ▶**Fig. 76.2.** Ear, nose, and throat (ENT) service demonstrated a mass in the upper nasopharynx using fiberoptic endoscopy of the nasal cavity.
5. Describe the findings on the scans.
6. Describe at least two surgical approaches for repair of the above lesion.

## Answers

1. *What are the potential causes of spontaneous cerebrospinal fluid (CSF) fistula?*
   - Spontaneous CSF fistulas may occur remotely following skull base head trauma in any area of persistent outpouching, or dural defect through a fracture site.
   - They may occur in the spine as part of a remote trauma or pseudomeningocele or a spontaneous dural sleeve rupture.
   - Congenital arachnoid cysts, spinal Tarlov or arachnoid cysts, or congenital encephaloceles may also chronically or acutely leak CSF.
   - CSF leaks have also been reported due to congenital defects in the middle ear.[1]
2. *Where are the potential sites of CSF leak?*
   - CSF leak may occur in any area with one of the above pathologies, but obviously occult leaks into the nasal cavity or from the spine into the epidural or lumbar fascial compartments may occur and may be difficult to detect.
3. *What other clinical syndromes might result from CSF fistula?*
   - CSF fistula might result in CSF infection as in this case. Any chronic communication between the environment and meningeal space may produce acute, recurrent, or chronic meningitis or other central nervous system (CNS) infections.
   - Low-pressure headache may also be attributable to a chronic CSF leak.[2]
   - Finally, superficial hemosiderosis has been reported following chronic CSF fistula into a traumatic pseudomeningocele with neovascularization and repeated hemorrhage. This could obviously present with mental deterioration, hearing, balance loss, and other cranial nerve deficits.[3,4]
4. *What would be the diagnostic tests indicated in this patient?*
   - In a patient where CSF fistula is suspected, an MRI scan of the complete neuraxis would be imperative. In some instances, this may not be diagnostic.[2]
   - A CT scan can be helpful to demonstrate bone defects. Cisternal infusion of contrast media followed by a thin-cut CT, might be able to demonstrate extra CNS flow of fluid.[2]
   - In rare instances, a nuclear medicine study with lumbar cisternal infusion of radionucleotide tracer may also be used to demonstrate a small or slow communication, though with less anatomic detail.[1]
5. *Describe the findings on the scans.*
   - The coronal T2-weighted MRI scan and the coronal reconstruction of the postcisternal infusion of contrast CT scan both show a defect in the ethmoidal bone of the frontal cranial fossa with herniation of tissue and CSF through the defect into the nasal cavity.
6. *Describe at least two surgical approaches for repair of the above lesion.*
   - There are essentially two surgical approaches to this lesion.
     - A bifrontal craniotomy with repair of the defect from above using bone, muscle graft, and a vascularized pericranial flap would be possible. This might be possible entirely with an extradural approach, but an intradural inlay graft might protect better against CSF leak.[1,2]
     - Alternatively, there is significant experience in some centers with an entirely endoscopic intranasal approach to the repair of this lesion with again vascularized mucosal flap and an inlay graft if possible.[5]

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# Case 77 Cerebral Palsy and Selective Dorsal Rhizotomies

*Jean-Pierre Farmer and Abdulrahman J. Sabbagh*

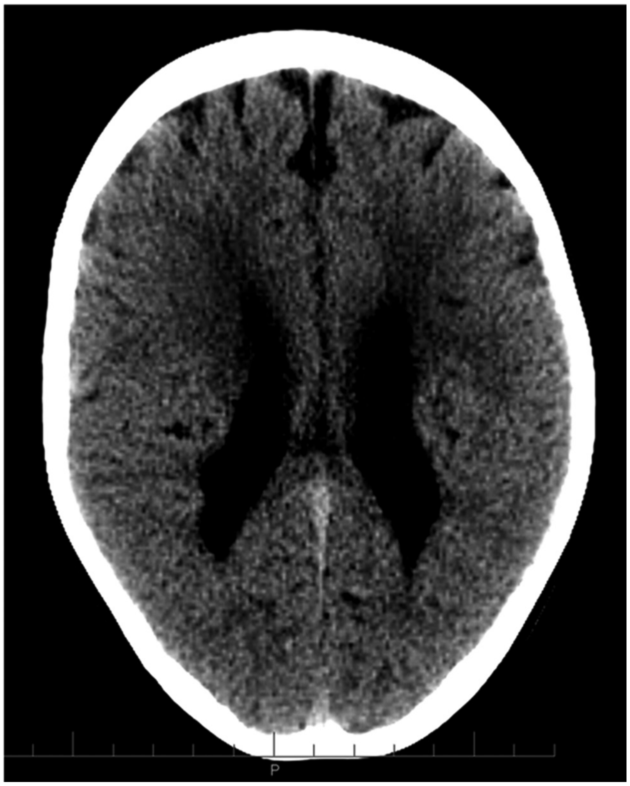

**Fig. 77.1** CT scan of the head showing presence of periventricular leukomalacia without hydrocephalus.

## Clinical Presentation

- A 3-year-old child is referred to you for surgical treatment of refractory spasticity.
- On examination, he is able to crawl in a reciprocal fashion predominantly and has started to use a walker.
- An earlier CT scan of the head is available (▶**Fig. 77.1**).

## Questions

1. What are your surgical treatment options?
2. What key features would you like to elicit in the history to determine that this young boy is a candidate for selective dorsal rhizotomy?
3. What physical features would make you think that he is a candidate for this procedure?
4. What features, if present in the history or physical examination, would make you rule against offering rhizotomies?
5. What factors would favor offering rhizotomies in your interpretation of the imaging?
6. What other investigations would you like to obtain on this child prior to planning rhizotomies?
7. What can you tell the family with respect to the potential outcome for the child with selective dorsal rhizotomies as opposed to continuing with physiotherapy and occupational therapy alone?
8. If there is involvement in the upper extremities, how would this influence your decision in terms of offering rhizotomies versus a baclofen pump?
9. What is the relative cost of the two treatment procedures over a lifetime?

## Questions (*continued*)

10. What are the risks and long-term sequelae following selective dorsal rhizotomies?
11. What would you recommend your anesthetist to use intraoperatively during stimulation?
12. What criteria would you use to determine your lesioning pattern?
13. What do you do to reduce the risk of sphincteric difficulties in these patients?
14. How do you control pain and spasticity perioperatively?
15. What adjuvant treatment will the child benefit from in the 2 months following the procedure?
16. What is the expected long-term outcome with respect to lower extremity function, activities of daily living, and upper extremity fine motor control following rhizotomies?
17. Is the preoperative status of the patient a strong determinant with respect to the long-term outcome of the child?

## Answers

1. *What are your surgical treatment options?*
   - Selective dorsal rhizotomy[1]
   - Baclofen pump placement[2,3]
2. *What key features would you like to elicit in the history to determine that this young boy is a candidate for selective dorsal rhizotomy?*
   - A history of prematurity, low birth weight, late acquisition of motor milestones, but progress being made.[1]
   - Other key points: The absence of previous surgical interventions, the absence of movement disorders other than spasticity, the absence of a family history involving neurologic diseases, and a good collaboration with therapists are all positive features.
3. *What physical features would make you think that he is a candidate for this procedure?*
   - Candidates should show evolving locomotor skills with adequate balance in the sitting position and good protective responses in the short leg sitting position.
   - Patient should show no associated movement disorder such as chorea or choreoathetosis and should show velocity-dependent increased tone and hyperreflexia with clonus, as well as limited range of motion.[1]
   - There should be adequate underlying strength with the squat-to-stand testing.
   - Patient should show at least quadruped falling or bunny hopping.
   - If he or she shows upright locomotor function, this should be even better.
4. *What features, if present in the history or physical examination, would make you rule against offering rhizotomies?*
   - Absolute contraindications include the presence of multiple orthopedic releasing procedures for short tendons, choreoathetosis, double hemiplegia, the inability to collaborate with therapists, associated significant cognitive difficulties, and dislocated hip.[4]
   - Relative contraindications include atypical perinatal history with birth at term, but with associated confounding factors such as meningitis, trauma in the neonatal period, or other prenatal factors.[4]
   - Hydrocephalus, if well dealt with, is not an absolute contraindication.
   - Other absolute contraindications would be severe motor restrictions or the presence of severe scoliosis preoperatively.
5. *What factors would favor offering rhizotomies in your interpretation of the imaging?*
   - Imaging of the brain with a CT scan or preferably MRI should show presence of periventricular leukomalacia (▶ **Fig. 77.1**) without hydrocephalus.
   - Presence of basal ganglia damage would be a relative contraindication.[1]
   - The spine X-rays should not show significant scoliosis.
   - The hip X-ray should show at least 50% or better femoral head coverage.
6. *What other investigations would you like to obtain on this child prior to planning rhizotomies?*
   - Other investigations would include the presence of gross motor measure scores, alignment scores, occupational therapy grading, and urodynamic testing.[5]
7. *What can you tell the family with respect to the potential outcome for the child with selective dorsal rhizotomies as opposed to continuing with physiotherapy and occupational therapy alone?*
   - The family has to be aware that randomized control trials have revealed an advantage of doing rhizotomy over simply continuing with physiotherapy and occupational therapy alone.
   - Abundant literature attests that there are persistent significant gains in gross motor function measure (GMFM), urodynamic profile, and the upper extremity, as well as activities of daily living gains, which are durable and substantial.[1]
8. *If there is involvement in the upper extremities, how would this influence your decision in terms of offering rhizotomies versus a baclofen pump?*
   - This decision remains controversial as both intrathecal baclofen pumps and rhizotomies have been used to treat upper extremity spasticity successfully.[1–3]

## Answers (*continued*)

- Because of the fact that rhizotomies have been shown to improve upper extremity function significantly, if a candidate meets the criteria for rhizotomies this may be regarded by some as the procedure of choice over baclofen pump placement.[1]
- Baclofen pump can provide effective long-term treatment of spasticity of cerebral origin, and its effects do not appear to diminish with time.[2]
- Intrathecal baclofen has been shown to reduce spasticity in the upper and lower extremities, and improve upper extremity function and activities of daily living.[3]
- However, baclofen pumps require frequent refills, change of battery packs, and repositioning of the catheters. They can occasionally produce drug toxicity, and the overall cost may be quite significant.[2,3]

9. *What is the relative cost of the two treatment procedures over a lifetime?*
   - The relative cost of the two procedures is about 4:1 (baclofen pump compared with rhizotomies).
   - Baclofen pumps remain a significantly more expensive way to treat spasticity.[1]
10. *What are the risks and long-term sequelae following selective dorsal rhizotomies?*
    - Long-term sequelae of selective dorsal rhizotomies are few if the procedure is done with stimulation and in a moderate lesioning fashion.
    - Reported problems include a higher incidence of scoliosis, which has to be compared with the natural history of the disease.[6]
    - In our experience, the rate of scoliosis is relatively high; however, the scoliosis curves are almost exclusively between 10 and 15 degrees, which present a debatably low clinical significance.[6]
    - Hyperlordosis and spinal stenosis have also been reported, although these are very rare. The clinical importance is again similar to the scoliosis cases, where cases of significance are the few ones who have undergone laminotomy or laminoplasty procedure.[6]
    - Bladder dysfunction is also a rare event.[5]
11. *What would you recommend your anesthetist to use intraoperatively during stimulation?*
    - During surgery, if done with stimulation, the anesthetist should use a combination of sufentanil or remifentanil at a low dose and propofol with nitrous oxide.
    - The neuromuscular junction transmission and the spinal cord transmission should not be altered by the anesthetic regimen during stimulation.[4]
12. *What criteria would you use to determine your lesioning pattern?*
    - The criterion most frequently used is that of spread to contralateral or upper extremity segments from stimulation of lumbosacral dorsal roots.
    - After-discharges and amplitude of stimulus response also play a role if spread is demonstrated to contralateral or upper extremity segments.[7]
13. *What do you do to reduce the risk of sphincteric difficulties in these patients?*
    - Care has to be taken to preserve S3 and S4 roots and to limit the lesioning at S2.
    - We tend to limit lesioning to 50% of both dorsal S2 roots combined, preserving at least one-third of S2 dorsal rootlets on each side.
    - With this pattern we have not identified long-term problems with bladder dysfunction and have even documented improvements in urodynamic profile.[1]
14. *How do you control pain and spasticity perioperatively?*
    - Postoperative pain is usually related to the length and depth of the incision, but also to some element of deafferentation as a result of the sectioning of the nerves.
    - An epidural catheter placed for the delivery of epidural morphine at T9–T10 that is above the surgical site is very helpful.[1]
    - In addition, oral diazepam can be beneficial but should be given at half of the recommended dose to avoid depressant effects on respiratory drive.
15. *What adjuvant treatment will the child benefit from in the 2 months following the procedure?*
    - Most centers will recommend enhanced physiotherapy and occupational therapy with the use of a pool therapy, arts and crafts, and the use of an exercise program to stretch resistant contractures, strengthen musculature, and date training.[1]
16. *What is the expected long-term outcome with respect to lower extremity function, activities of daily living, and upper extremity fine motor control following rhizotomies?*
    - Lower extremity function using objective measurement such as the GMFM score and alignment scores shows durable improvements, as do activities of daily living and upper extremity fine motor control following rhizotomies.[4,8,9]
    - These improvements have been found to last up to 5 years postoperatively and still improving at that point.[8]
17. *Is the preoperative status of the patient a strong determinant with respect to the long-term outcome of the child?*
    - Yes, the better the patient is preoperatively, the more favorable the outcome will be.
    - In more favorable cases, gains will bring the patient closer to the normal age-matched controls with respect to motor and upper extremity function.[1]

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# Case 78 Neural Tube Defect

*Abdulrahman J. Sabbagh, Abdulrahman Y. Alturki, José Luis Montes, Jean-Pierre Farmer, and Jeffrey Atkinson*

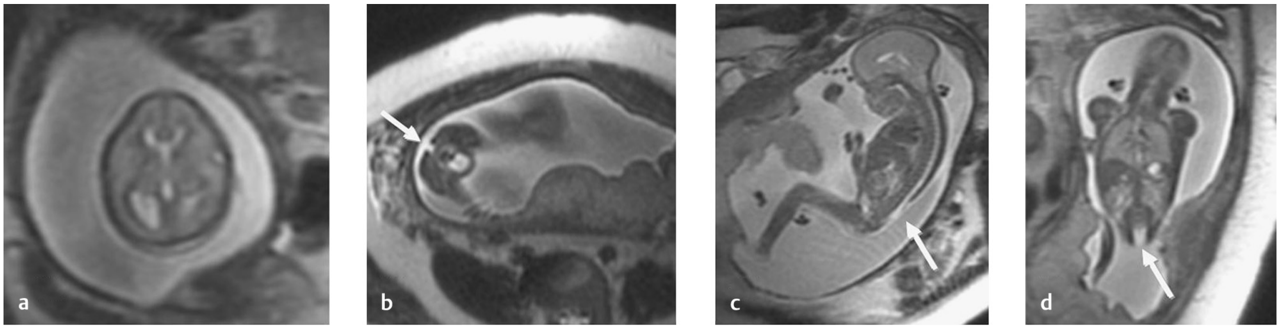


**Fig. 78.1** Prenatal MRI scans with (**a**) T2-weighted axial and (**b**) sagittal sections of the brain in utero. (**c**) T2-weighted sagittal and (**d**) coronal sections of the spine are also shown.

## Clinical Presentation

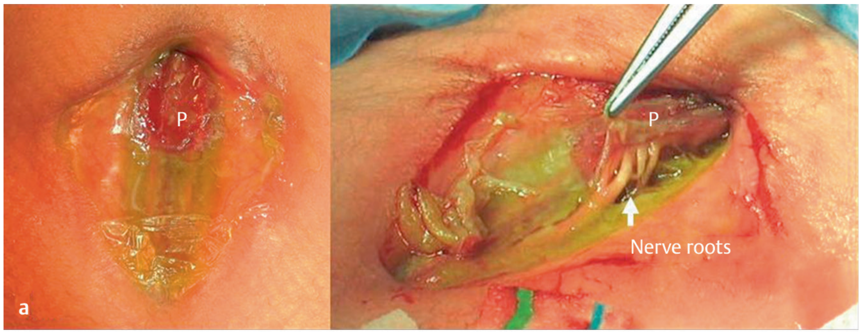
- A 32-year-old woman is referred to you from an obstetrician. She is 22 weeks pregnant, gravida 2 para 1 (G2P1) but otherwise healthy with a normal first daughter.
- A uterine ultrasound was done followed by a fetal MRI scan (▶**Fig. 78.1**).
- She has a cesarean section and a lesion is found in the midline on her infant's back.
- An MRI of the infant is done in the first few hours of its life (▶**Fig. 78.2**).

## Questions

1. What is the diagnosis? Describe the MRI shown in ▶**Fig. 78.1**.
2. Give five features on this MRI of the brain and spinal cord that are characteristic of this diagnosis. Why is there pneumocephalus and air in the spinal canal?
3. Describe methods of evaluation for this condition in the prenatal period.
4. What are the most common sites for neural tube defects (NTDs)?
5. How would you assess the level of the defect clinically?
6. What do you tell the parents regarding survival (with or without treatment), intelligence, ambulation, and urinary continence? What are the causes of early and late mortality?
7. Highlight a management plan.
8. When should you operate on this patient? What are the goals and principles of the surgery?
9. Describe surgical options for repair of such large defects in the skin.
10. What are the risk factors associated with NTDs?
11. How can NTDs be prevented?

## Answers

1. *What is the diagnosis? Describe the MRI shown in* ▶**Fig. 78.1**.
   - The diagnosis is of Chiari II malformation with an open myelomeningocele (MMC).
   - The prenatal MRI shows a T2-weighted axial section of the brain in utero depicting asymmetrical ventriculomegaly. The axial section at the lumbar level shows an open MMC (arrow). Sagittal and coronal sections show evidence of the MMC and Chiari II.
2. *Give five features on this MRI of the brain and spinal cord that are characteristic of this diagnosis. Why is there pneumocephalus and air in the spinal canal?*
   - Characteristic findings include the following[1]:
     - Medullary kinking
     - Tectal beaking
     - Enlarged massa intermedia
     - Elongation and/or cervicalization of the medulla
     - Low attachment of tentorium



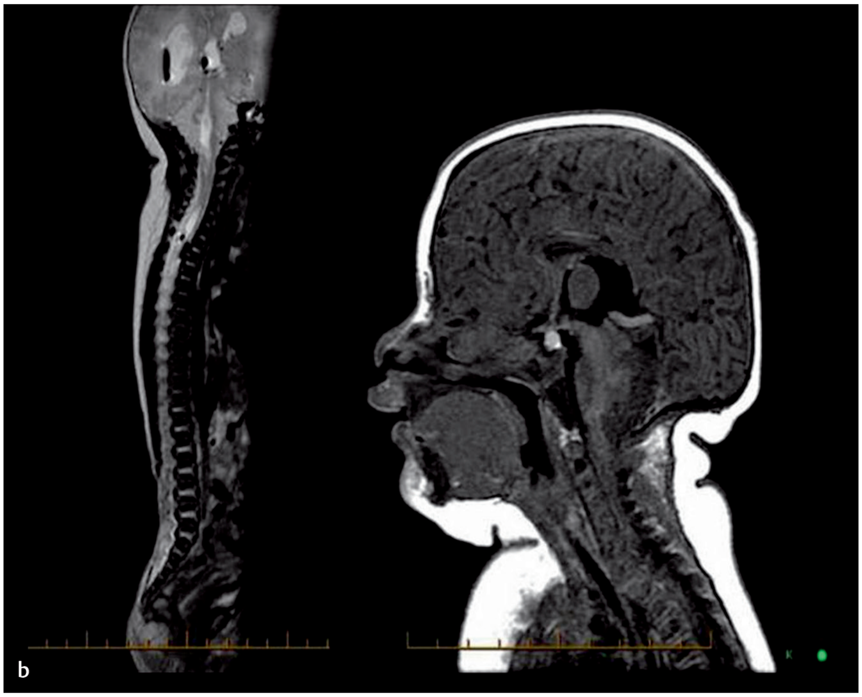


**Fig. 78.2** (**a**) Preoperative and intraoperative photograph of the skin defect depicting neural placode (P) and nerve roots. **(b)** MRI scans obtained shortly after birth with sagittal T2-weighted image of the spine and T1-weighted image of the brain.

## Answers (*continued*)

  - Hydrocephalus
  - Syringomyelia in the area of the cervicomedullary junction
  - Dysgenesis of the corpus callosum
- Pneumocephalus and air in the spinal canal indicate open MMC.

3. *Describe methods of evaluation for this condition in the prenatal period.*
   - Methods of evaluation include:
     - Mother serum alpha-fetoprotein (AFP)
     - Prenatal ultrasound (high NTDs)
     - If both are positive, then an amniocentesis may be performed for amniotic AFP and acetylcholinesterase levels (the diagnostic accuracy for NTD rises to more than 97%).[2]
     - Fetal MRI is an option when available, as it gives accurate information regarding associated anomalies and may help in prognostication.
4. *What are the most common sites for neural tube defects (NTDs)?*
   - Most common sites of occurrence include the lumbosacral area in 50% of cases, followed by the thoracolumbar area in 35% of cases.[3]
5. *How would you assess the level of the defect clinically?*
   - The defect level is determined by assessing the lowest level of neurologic function (▶**Table 78.1**).[4]
6. *What do you tell the parents regarding survival (with or without treatment), intelligence, ambulation, and urinary continence? What are the causes of early and late mortality?*
   - Prognosis is described below.[5–7]
     - Survival
       - ○ Without treatment, only 14–30% patients survive infancy.
       - ○ With treatment, 85% survive.
     - Intelligence
       - ○ Seventy to 80% will have normal intelligence quotients.
       - ○ Mental delays may be related to shunt malfunctions, primary microgyria, or absence of corpus callosum.

## Answers (*continued*)

- Ambulation
  - Fifty percent are ambulatory.
  - Up to 80% can ambulate with bracing. However, 80% of these will decrease over time because of weight gain by the patients.
- Urinary incontinence
  - Ninety to 95% are incontinent but are able to stay dry with intermittent catheterizations.
- Mortality
  - Early mortality is related to Chiari malformation complications such as aspiration and respiratory arrest.
  - Late mortality may be related to shunt complications, urosepsis, as well as progressive respiratory compromise due to kyphoscoliosis.

7. *Highlight a management plan.*
   - A management plan is described below.
     - Items related to Chiari II malformation[1]:
       - Measure head circumference to follow the rate of growth due to risk of hydrocephalus.
       - Obtain a head ultrasound within 24 hours of birth.
       - Check for inspiratory stridor and apneic episodes.
     - Items related to the defect[8]:
       - Measure the size of the defect.
       - If the lesion is ruptured, start antibiotics.
       - Cover the lesion with a piece of sterile Telfa (Tyco Healthcare, Mansfield, MA).
       - Keep the patient in Trendelenburg position (to keep pressure off lesion).
       - Plan early surgical closure.
     - General assessment and management:
       - Neonatologist assessment for other abnormalities
       - Urologic consultation and regular urinary catheterization
       - Orthopedic consultation for spine, hip, and knee deformities
8. *When should you operate on this patient? What are the goals and principles of the surgery?*
   - Timing of surgery should be within 48 to 72 hours.[6, 9]
   - The goals of surgical repair include reconstruction of the neural tube and its coverings, avoidance of meningitis, and protection of the remaining functional tissue in the neural placode.
   - The principles of the surgery include:
     - Reconstruction of the neural tube (▶**Fig. 78.3**a)
       - The placode is dissected from the surrounding tissue by incising the junctional zone.
       - All dermal remnants are resected, and the neural tube is reconstituted by closing the pia with a 7–0 monofilament suture.
     - Reconstruction of the thecal sac (▶**Fig. 78.3**b)
       - The dura is dissected free from its junctions with the fascia and skin.
       - The goal is a watertight closure without causing constriction of the closed neural placode.
     - Often the defect is large enough to require a dural patch (▶**Fig. 78.3**c).
       - Tissue sealant is preferably used at the end of dural closure.
       - Midline fascia and skin closure in layers (▶**Fig. 78.3**d)
9. *Describe surgical options for repair of such large defects in the skin.*
   - Several options are available and usually best done by a plastic surgeon. They include:
     - Circumferential skin release dissection (used in small- to medium-sized defects)
     - Ramirez procedure: medial advancement of bilateral bipedicled musculocutaneous flap based on the latissimus dorsi and maximus gluteus without any relaxing incisions or skin grafting.[10]
     - Flaps used include latissimus dorsi myocutaneous flap for thoracolumbar defects and gluteus maximus myocutaneous flap for lower defects.[11]
     - The junctional zone or the cyst wall membrane can be used as a graft.
     - Alternatively, artificial dermis can be used.[12]
10. *What are the risk factors associated with NTDs?*
    - Risk factors include the following (the associated percentage risk of having a child with NTD is in brackets)[2]:
      - Partner diagnosed with NTDs (2–3% risk of having a child with NTD)
      - Previous pregnancy with NTDs (2–3%)
      - Epileptic on carbamazepine or valproic acid (1%)
      - Diabetes mellitus type I (1%)
      - Close relative with NTDs (0.3–1%)
      - Pregnancy with obesity and weight over 110 kg (0.2%)
      - Other risk factors include exposure to radiation, pesticides, anesthetic agents, hot tubs, smoking, and occupational hazards such as nursing or part of operating room staff (exposure to anesthetic agents).
11. *How can NTDs be prevented?*
    - Prevention can be achieved by supplementing expecting mothers with folic acid.
      - Expecting mothers with no known risk factors should take folic acid at the dose of 0.4 mg/day.
      - Those with a higher risk factor should take folic acid at 4 mg/day (for the duration of 1 month before pregnancy through the first trimester).[13]

**Table 78.1** Level of neurologic function and associated deficit

| Paralysis Below | Finding |
|---|---|
| T12 | Complete paralysis of all muscles in the lower limbs |
| L1 | Weak to moderate hip flexion, palpable contraction in sartorius |
| L2 | Strong hip flexion and moderate hip adduction |
| L3 | Normal hip adduction and almost normal knee extension |
| L4 | Normal hip adduction, knee extension, and dorsiflexion/inversion of the foot, some hip abduction in flexion |
| L5 | Normal adduction, flexion, and lateral rotation of the hip, normal knee extension, moderate flexion, normal foot dorsiflexion, hip extension absent; produces dorsiflexed foot and flexed thigh |
| S1 | Normal hip flexion and abduction/adduction, moderate extension and lateral rotation; strong knee flexion and inversion/eversion of the foot, moderate plantar flexion of the foot; extension of all toes, but flexion only of terminal phalanx of the great toe; normal medial and lateral hip rotation; complete paralysis of foot intrinsic muscles (except abductor and flexor hallucis brevis); produces clawing of toes and flattening of the sole of the foot |
| S2 | Difficult to detect abnormality clinically; with growth, this produces clawing of the toes due to weakness of intrinsic muscles of the sole of the foot (innervated by S3) |

*Source:* Sharrard WJ. The segmental innervation of the lower limb muscles in man. Ann R Coll Surg Engl 1964;35:106–122.

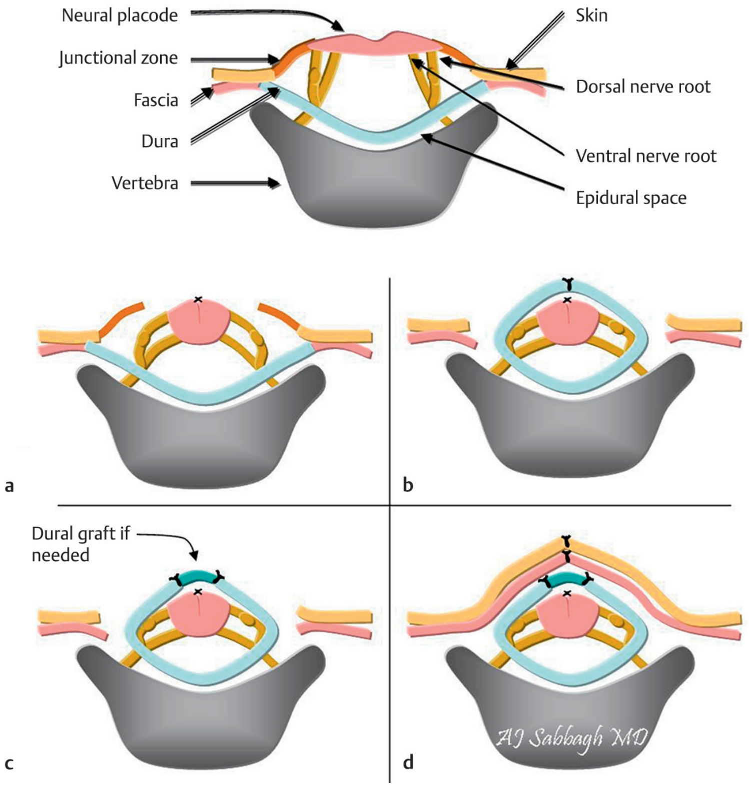


**Fig. 78.3** Artist's rendering of spinal dysraphism closure. See text for detailed description. Depicted steps include (**a**) reconstruction of the neural tube, (**b**) reconstruction of the thecal sac, (**c**) dural patching of a large defect, and (**d**) midline fascia and skin closure in layers.

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# 79 Idiopathic Syringomyelia in Children and Adolescents

*Nabeel S. Alshafai, Ivona Nemeiko, and Paul Steinbok*

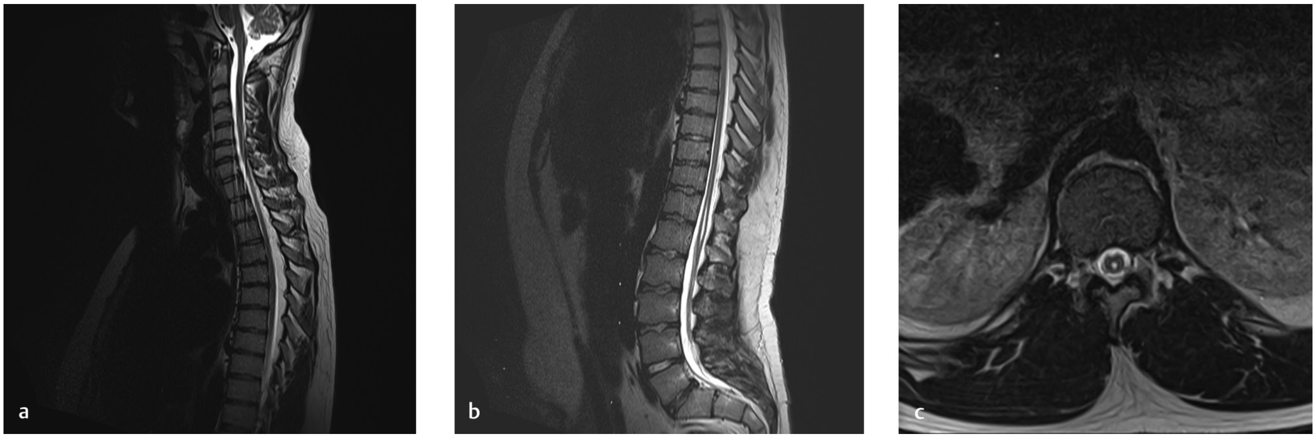

**Fig. 79.1** (**a**) MRI scan of the caudal posterior fossa and cervicothoracic spine in the sagittal plane. (**b**) MRI scan of the thoracolumbar spine in sagittal plane with visualized syrinx. (**c**) MRI scan of the caudal thoracic spine in axial plane on the level of syrinx.

## Clinical Presentation

- A 14-year-old boy was referred to our hospital for scoliosis and mild claw foot.
- No other abnormalities on the clinical examination were found.
- His most recent MRI is provided (see ▶**Fig. 79.1**).

## Questions

1. Describe your findings on the MRI scan. Which MRI sequences do you use? Is intravenous contrast always necessary? What other areas would you require to image?
2. What is the differential diagnosis for this lesion? What are the differential diagnoses for an intramedullary cystic lesion?
3. Classify syringomyelia?
4. Describe the differences between syringomyelia associated with Chiari malformation type 1 (CM1) and idiopathic syringomyelia (IS). Is there an age-related difference between the two conditions?
5. What is the prevalence of IS?
6. In IS, do symptoms correlate with the anatomical localization of the syrinx?
7. Which management options for IS do you know?
8. What is the prognosis for conservative management of IS?
9. What are the surgical indications for IS?
10. How would you manage this patient?
11. The patient has scoliosis. If the scoliosis is severe enough to require orthopaedic intervention, does the syrinx size change after the spinal fusion?

## Answers

1. *Describe your findings on the MRI scan. Which MRI sequences do you use. Is intravenous contrast always necessary? What other areas would you require to image?*
    - The MRI scan shows sagittal and axial T2-weighted image sequences of the spine. It illustrates a centrally located fluid filled cavity at the thoracolumbar segment that measures 2.4 mm in its maximum diameter. There is no mass or infiltrative lesion and there is no CM. The imaging characteristics are consistent with terminal syrinx or terminal hydromyelia.
    - The T2-weighted sequence is the most sensitive to diagnose syrinx/hydromyelia. Intradural intramedullary spinal tumors are important to exclude in the differential diagnosis, and if the scan is suggestive of a mass lesion, intravenous contrast injection is indicated. Recent studies[1] suggest that T2-weighted sequences are sufficient to exclude tumor when MRI (1.5 to 3T) is used even in the presence of irregular and thick septations within the syrinx. T2-weighted MRI is associated with very high sensitivity and 100% negative predictive value.
    - Brain MRI is not required especially in this case of terminal hydromyelia, which is often considered a normal variant of no clinical significance. Notice that brain MRI is indicated if the syrinx is in the cervical or cervicothoracic region, where hydrocephalus and/or other pathologies in the brain need to be ruled out. Visualizing the foramen magnum (FM) region is necessary to exclude potential obstruction in the cerebrospinal fluid (CSF) pathways as in CM.[2,3]
    - If the pathology is located more caudally like in this case, tethered cord should be excluded. Noninvasive MRI around the spinal syrinx using cardiac gated phase contrast CSF flow studies could be used to reveal CSF blockage and should be considered only in unusual cases with progressive symptoms, where surgical intervention is being considered.
2. *What is the differential diagnosis for this lesion? What are the differential diagnoses for an intramedullary cystic lesion?*
    - There is no specific differential diagnosis for the visualized lesion, which is pathognomonic as a case of ventriculus terminalis.
    - Differential diagnosis of the cystic lesion in the cord includes:
        - Cord neoplasms with cystic component
        - Myelomalacia
        - Persistent central canal/hydromyelia (ventriculus terminalis)
        - Primary and secondary syringomyelia
        - IS
3. *Classify syringomyelia?*
    - Syringomyelia is classified into primary, secondary, and idiopathic.
        - *Primary* syringomyelia is seen in younger patients and often associated with congenital malformations such as CM, myelomeningocele, Dandy–Walker or Klippel–Feil syndrome etc.
        - *Secondary* syringomyelia is observed in cases with previous trauma, spinal cord tumor, or arachnoiditis.
        - *Idiopathic* syringomyelia (IS) is seen in cases where no underlying cause is found. We often do not perform electrophysiological monitoring for IS.
    - The term “hydromyelia” is used to indicate distension of the residual central canal. If the fluid cavity dissects outside the central canal, then this represents a true syringomyelia. Often one observes a combination of both findings; hence the term hydrosyringomyelia is appropriate to describe such a finding. If the fluid is in the center of the cord, although the term “syrinx” is still considered correct nomenclature, strictly speaking this entity is actually hydromyelia. Hydromyelia almost never causes or presents with a neurologic deficit.[4]
    - Anatomical localization encompasses cervical, thoracic, lumbar, terminus and multilevel.
4. *Describe the differences between syringomyelia associated with Chiari malformation type 1 (CM1) and idiopathic syringomyelia (IS). Is there an age-related difference between the two conditions?*
    - The answer is summarized in ▶**Table 79.1**.
    - The vast majority of children with IS are asymptomatic or have symptoms, such as back pain that may not be related to the IS. Neurological findings are rare and most often include:
        - Gait or balance disturbance
        - Focal weakness
        - Hypo-/hyperreflexia
        - Bowel/bladder dysfunction
    - Sensory deficit[5,6,7,8,9]
        - In syringomyelia associated with CM1, there may be symptoms and signs referable to the tight FM or symptoms/signs related specifically to the syringomyelia.
        - The size of the syrinx in IS is usually smaller than in CM1 and no descent of tonsils occurs in IS.
        - One study suggested a hypothesis of similarities of IS with CM1 in *adults* and found that both pathologies are coexisting with small posterior fossa.
        - The differences are summarized in ▶**Table 79.2.**
5. *What is the prevalence of IS?*
    - Syringomyelia is a quite rare disease. The prevalence for syringomyelia of all etiologies varies from 1.9 per 100,000 (Japan) up to 8.2 to 8.4 per 100,000 in the

**Table 79.1** Clinicoradiological features of syrinx in IS at all ages and when compared to CM1

| | **Pediatric (< 19 yr old)** | | **Adults** | |
|---|---|---|---|---|
| | **CM 1** | **IS** | **CM 1** | **IS** |
| **Etiological distribution** | 41–79%[8] | 6.2%[8] | 28–47%[8] | 17–24% (> 70 yr old 38%)[8] |
| **Clinical presentation** | | | | |
| Pain (back/neck/lower extremities/headache) | **Common** | **Sometimes** | **Sometimes** | **Common** |
| Scoliosis | **Common** | **Sometimes** | **Sometimes** | **Sometimes** |
| Neurological findings (motor, sensory, balance, gait impairment) | **Common** | **Sometimes** | **Common** | **Common** |
| Others | **Sometimes**<br>*Craniovertebral junction abnormalities (basilar invagination, platybasia, small posterior fossa, concavity in clivus, occipitalization of atlas, spina bifida in upper cervical region), spinal dysraphism and tethered cord*[29] | **Rare**<br>*Developmental anomaly or cutaneous markers fx. hemangioma, midline hair patches in 12.5%* | **Rare** | **No** |
| *References to clinical presentation* | 13,29,31,35,38,40,42,44 | 18,19 | 17,32,26,32,33,43,49 | 17.26,27 |
| **Radiological features** | | | | |
| Syrinx levels | Cervical<br>Cervicothoracic | Thoracic<br>Cervicothoracic<br>Cervical | Cervicothoracic<br>Cervical | Thoracic<br>Cervicothoracic |
| Tonsils descent (≥5 mm below FM) | + | – | + | – |
| Communications with fourth ventricle | No | No | No | No |
| Coexisting hydrocephalus | Rare | – | Rare | – |
| CSF flow obstruction at FM | + | –[a] | + | – |
| Syrinx diameter in mm (most of the time) | >5 | <5 | >5 | <5 |
| References to radiological features | 29,40 | 18,19,24,29,54 | 20,26,47,50 | 17,20,25,26,27 |

*Abbreviation:* CM1, Chiari malformation type 1; CSF, cerebrospinal fluid; FM, foramen magnum; IS, idiopathic syringomyelia.
[a] Nomenclature unclearness; in IS cases, no CSF flow obstruction can be found.
Etiological distribution among all MRI verified various syringes in pediatric and adult patient groups

**Table 79.2** Radiological features of syrinx in IS compared to CM1 in adults

| **Imaging Findings** | **IS** | **CM1** |
|---|---|---|
| Tonsils descended | – | ↓ ↓ |
| Diameter of fossa posterior | ↓ | ↓ |
| Ventral CSF space | ↓ | ↓ |
| Dorsal CSF space | ↓ | – |
| Syrinx diameter > 3 mm | ↑ ↑ | –/ ↑ |

*Abbreviations:* –, absent; ↓, decreased; ↑, increased; ↓ ↓, significantly descended; ↑ ↑, very often present.

## Answers (*continued*)

western countries' population.[10] Among all the above, idiopathic etiology has been found only in 6.2% in pediatric patients and 20 to 30% in adults. The number of diagnosed syringomyelias is increasing.

6. *In IS, do symptoms correlate with the anatomical localization of the syrinx?*
   - A clear correlation between IS pain symptoms and anatomical localization of the syrinx has not been observed in all age groups.
   - Rodriguez et al,[5] observed only 39% concordance between the clinical course of IS and syrinx size in neuroimaging. They followed the pediatric IS subgroup of patients who were treated conservatively. Interestingly, 40% improved clinically despite no radiological changes.
7. *Which management options for IS do you know?*
   - The answer is summarized in ▶**Table 79.3**.
     - Conservative treatment
       Conservative management in the form of observation and clinicoradiological follow-up is the most common practice as IS is a benign condition.
     - Surgical treatment
       - Used very rarely.
       - Doing any myelotomy in a small syrinx or hydromyelia is contraindicated and much more likely to have a harmful effect. The procedure with shunting should be reserved only for IS cases with large syringes and only when the patient is deteriorating and no other cause can be found.
       - A CSF flow study with MRI cardiac gate imaging is reasonable to do if one is considering the need for surgery. If one can demonstrate a block to CSF flow, then one may consider a decompression with lysis of adhesion at that level (a very rare situation).
       - Other described surgical methods are on a rare case report basis (See ▶**Table 79.3**).
8. *What is the prognosis for conservative management of IS?*
   - When followed for a period of up to 12 years in pediatric cases and 20 years in adults, most patients with IS remain stable or even improve clinically except for < 10%, who show signs of clinical deterioration. On follow-up imaging, the syrinx size remains unchanged except in 20% cases. In these 20% patients, approximately half of their syrinx increases and the other half decreases in size unrelated to their clinical presentation, which often remain unchanged or improve. The detailed answer is visualized in ▶**Table 79.4.**
9. *What are the surgical indications for IS?*
   - There is rarely an indication for surgical treatment in IS. Most patients were operated only in cases with neurological deterioration. Clinical improvement after the surgery, is variable, attesting to the concern that symptoms are not necessarily related to the identified IS. This is all the more reason to avoid surgery as much as possible.[5,6]
   - In reviewing the literature, the two largest series of pediatric IS had only one case operated by syringo-subarachnoid shunt and another by fenestration. However, these surgical solutions were not associated with significant clinical improvement in one patient and a clinical deterioration in the other[6] (see ▶**Table 79.3**).
10. *How would you manage this patient?*
    - The management in the presented case is conservative management with clinical follow-up. *Repeat MRI is not needed if the patient is stable.* Spontaneous resolution of syrinx or clinical symptoms improvement can be expected.[5,6,8]
11. *The patient has scoliosis. If the scoliosis is severe enough to require orthopaedic intervention, does the syrinx size change after the spinal fusion?*
    - The syrinx size after the spinal fusion remains unchanged in most of the cases. The surgery can be successfully done with the use of perioperative neuromonitoring. No increase in complication rate for scoliosis surgery was found between groups with normal and abnormal MRI.

**Table 79.3** Pediatrics (< 19 yr old) patients with IS

| Studies | Total No. of patients in the study | Conservative | Syringosubarachnoidal shunt | Fenestration of the syrinx and sectioning of the FT | CSF withdraw (lumbar puncture) | Posterior fossa decompression (suboccipital craniectomy) |
|---|---|---|---|---|---|---|
| **Magge[6] (2011)**<br>(No. of patients) | 48 | 46 | 1 | 1 | 0 | 0 |
| **Rodriguez[5] (2015)**<br>(No. of patients) | 98 | 98 | 0 | 0 | 0 | 0 |
| **Joseph[9] (2013)**<br>(No. of patients) | 39 | 35 | 0 | 0 | 4 | 0 |
| ***Clinicoradiological follow-up period*** | | | | | | |
| **Magge[6] (2011)** | | 1–84 mo | 4 yr | Lack of information | None | None |
| **Rodriguez[5] (2015)** | | 1–143 m | None | Lack of information | None | None |
| **Joseph[9] (2013)** | | 4–84 mo | None | None | 16–18 mo | None |
| ***Outcome*** | | | | | | |
| **Clinical** | | 80% unchanged/improved[18,19] | New weakness in lower extremities, unsteady gait | No change in symptoms and clinical status | All improved in the back pain in FU; recurrence in 1 case (18 months) | Used as a surgical treatment in some cases with radiological features of Chiari 0 malformation[a] |
| **Radiological (syrinx size)** | | 80% stable | decrease in syrinx size 7 → 4 mm | stable syrinx size 8.5 mm | unchanged | |

*Abbreviations:* CSF, cerebrospinal fluid; FT, filum terminale

[a] Chiari 0 malformation term is used to describe the clinical status with no tonsil descent, but still compromise in CSF flow at the foramen magnum in radiological imaging.[25] There seems to be nomenclature differences. In some papers, IS terminology is used despite radiological findings with CSF flow compromise at the level of the foramen magnum.[24]

**Table 79.4** Course of the conservative treatment of syringomyelia in pediatric patients (<19 yr old)

| Outcomes | CM1 | IS |
|---|---|---|
| *Clinical* | | |
| Follow-up | 12 mo–20 yr | 1 mo–12 yr |
| Scoliosis, pain, neurological status, asymptomatic | **Stable or improved**[30,33]<br>(up to 37% worsened)[35,39] | **Stable or improved**[18,19,54]<br>(up to 10% worsened) |
| *Radiological* | | |
| Follow-up | 1 mo–16 yr | 2 mo–7 yr |
| Syrinx size | **Mostly unchanged**[34,53] | **Unchanged**[18,19]<br>(13% increased vs. 8% decreased) |

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# Case 80 Tethered Cord Syndrome

Saleh S. Baeesa

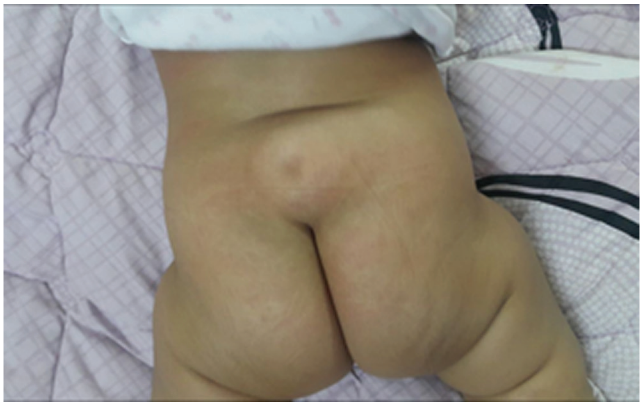

**Fig. 80.1** A 20 × 25 mm midline lumbar swelling covered with normal skin.

## Clinical Presentation

- A 6-month-old girl is brought by his parents to the outpatient clinic with a history of lower back swelling since birth.
- She is their first baby, which was a product of an uneventful pregnancy and was delivered at full-term through a vaginal route.
- At birth, a normal-looking baby was observed with a head circumference at the 50th percentile for age; a soft 20 × 25 mm midline lumbar swelling covered with normal skin was seen (▶**Fig. 80.1**).
- The neurological examination was within normal limits, in particular spontaneous lower extremities movements were seen as well as movements in response to cutaneous stimuli. There was normal anal shape and reflex.

## Questions

1. Interpret the MRI findings (▶**Fig. 80.2**, ▶**Fig. 80.3**).
2. What is the most likely diagnosis?
3. How would you anatomically classify this condition?
4. Describe the embryogenesis of this anomaly.
5. Describe the symptoms that may arise due to tethering of the spinal cord.
6. Describe the pathophysiology of tethered cord syndrome (TCS).
7. Describe the timing and the rationale of surgical treatment of asymptomatic children with TCS.
8. Describe the utility of intraoperative monitoring.
9. Briefly outline the technical steps performed during the surgical procedure (▶**Fig. 80.4**).
10. Following an untethering surgery with resection of the lipoma, the child develops a CSF leak. Outline the different options available for management of this complication.

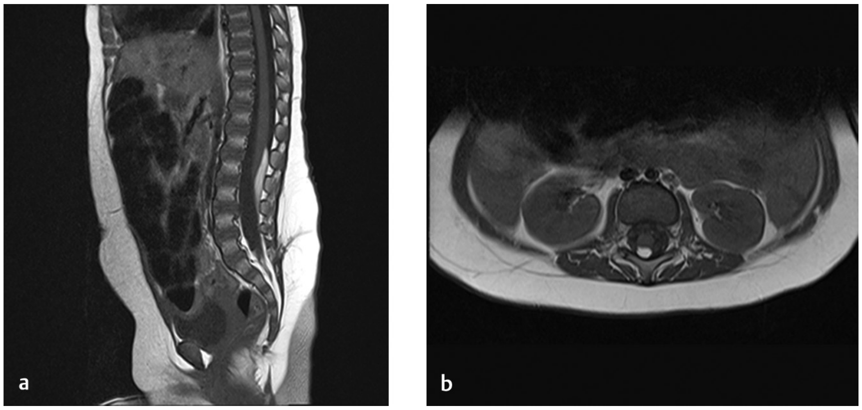

**Fig. 80.2** Lumbar spine sagittal T1-weighted MRI image (**a**) and axial T1-weighted MRI image of above described newborn (**b**).

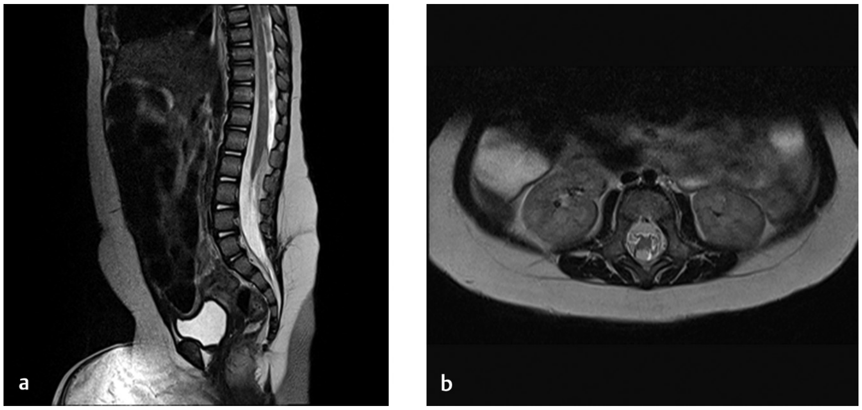

**Fig. 80.3** Lumbar spine sagittal T2-weighted MRI image (**a**) and axial T2-weighted MRI image of above described newborn (**b**).

## Answers

1. *Interpret the MRI findings* (▸**Fig. 80.2**, ▸**Fig. 80.3**).
    - The sagittal T1-weighted MRI image (▸**Fig. 80.2**) revealed a hyperintense subcutaneous lumbosacral swelling with central hypointense (fibrous) bundles extending to an S1 lamina defect which extend intradurally upward to be fused with the dorsal surface of the conus of the spinal cord. The lesion is isointense on T2-wieghted images (▸**Fig. 80.3**). The spinal cord ends at L2–L3 level without associated cord abnormalities.
2. *What is the most likely diagnosis?*
    - Spinal cord lipoma.
3. *How would you anatomically classify this condition?* The anatomical classification of spinal cord lipoma can be as follows:
    - Dorsal lipoma: located entirely along the dorsal surface of the spinal cord and does not contain nerve roots. The lipomatous tissue extends through the dural defect to blend with the extraspinal subcutaneous fat. There is associated thickened filum terminale that extends from the lower end of the conus.
    - Transitional lipoma: similar to the dorsal lipoma, but usually larger with wider lipomatous tissue–cord junction (fusion line) with involvement of the lower end (tip) of the conus, which may be rotated to one side. The nerve roots are found exiting lateral and ventral to the fusion line and there may or may not be a thickened filum. The lipoma extends through a larger dural defect to the extraspinal subcutaneous tissue.
    - Terminal lipoma: unlike the above two types, it attaches to the caudal end of the (normal looking) conus with the sacral roots exiting rostral to the lipoma. There may be a thickened short filum anchoring the conus to the lipoma or the lipoma may replace the filum. The dura and its musculofascial coverings are intact.
    - Chaotic lipoma: it may start as dorsal, transitional, or terminal types but it extends caudally to be ventral to the neural tissue and engulf the lumbosacral roots. There is no clear demarcation between the lipomatous tissue and neural tissue. There may be an associated sacral agenesis.
4. *Describe the embryogenesis of this anomaly*.
    - The embryologic defect present in the pathogenesis of dorsal and transitional spinal cord lipomas occurs during primary neurulation: there is mesodermal invasion of the neural tube due to premature disjunction between the cutaneous and neural ectoderms, before fusion of the neural tube. Chaotic lipomas, on other hand, are associated with a different embryologic defect. Their high association (up to 80%) with caudal agenesis suggests that these malformations originate from a general

## Answers (*continued*)

failure of caudal cell mass during the early stages of secondary neurulation.

5. *Describe the symptoms that may arise due to tethering of the spinal cord.*
    - Children with spinal cord lipomas can have variable presentation from being asymptomatic (presenting only with a lumbar mass) to having evident neurological, urological, or orthopedic abnormalities as a result of spinal cord tethering. These abnormalities can manifest in the form of symmetric or asymmetric lower extremity weakness, loss of sphincter function, scoliosis, and joint deformity.
6. *Describe the pathophysiology of tethered cord syndrome (TCS).*
    There are multiple mechanisms explaining the pathophysiology of TCS. Correlations between reported experimental results and clinical observations have been described as follows:
    - Spinal cord tethering associated with stretching produces excessive tension within the cord tissue and can result in metabolic derangements which can lead to histological changes.
    - Impairment of oxidative metabolism can be proved by the various degrees of reduced state of cytochrome a, a3, which indicates slow electron transport coupled with adenosine diphosphate (ADP) phosphorylation or slow adenosine triphosphate (ATP) use.
    - Oxidative metabolism is impaired in the spinal cord under steady traction and the severity of neuronal dysfunction parallels the degree of impairment in oxidative metabolism.
    - A decrease in blood flow and glucose metabolic impairment is correlated with cytochrome reduction. Increase in blood flow is reported to be associated with postsurgical neurological improvement.
    - Chronic cord traction may be compensated for by the plasticity of the spinal cord tissues. Spontaneous alleviation or fluctuation of a patient's symptoms may be explained by the neuronal plasticity. Symptoms tend to recur or progress as a result of repeated cord stretching due to spine flexion and extension, as well as lateral flexion.
    - Irreversible neuronal damage can result from sudden stretching of the already chronically tethered cord. TCS patients who have no satisfactory surgical results may correspond to this experimental model of cord tethering. Long tract signs in human TCS may be caused by similar mechanisms.
7. *Describe the timing and the rationale of surgical treatment of asymptomatic children with TCS.*
    - There is clear evidence that TCS is caused by steady excessive tension in the lumbosacral cord accentuated by repeated stretching of the spinal cord by flexion or extension movements. Histologically, demonstrable neuronal damage is associated with sudden additional violent stretching force applied to the cord, which may explain the irreversible neurological deficits in some TCS patients. Early untethering of the spinal cord is advisable in TCS patients in general. Therefore, children with symptomatic (TCS) and spinal cord lipomas usually undergo surgical untethering as early as possible. Surgical treatment of asymptomatic spinal cord lipomas remains controversial. Given that children have an actuarial lifespan of around 70 years worldwide and given that established neurogenic bladder and neurological deficit seldom recover after untethering, prophylactic untethering becomes an attractive option when it offers a better outcome than conservative therapy; one should also take into account individual center outcomes and operator experience when deciding whether or not to operate.
8. *Describe the utility of intraoperative monitoring.*
    - Intraoperative electrophysiological monitoring is the sine qua non in spinal cord lipoma surgery. A standard electromyography (EMG) recording is performed with needles inserted into the rectus femoris (L4), tibialis anterior (L4–L5), gastrocnemius (S1), and abductor halluces muscles bilaterally. Small-gauge needles are inserted bilaterally at the level of the external anal sphincter. This can be combined with transcortical motor evoked potentials, which are recorded from the electrodes at the external anal sphincter level.
9. Briefly outline the technical steps performed during the surgical procedure (▶**Fig. 80.4**).
    - Surgical steps include:
        - Exposure and dissection of the extraspinal lipomatous stalk
        - Detachment of the lipoma from the dural defect
        - Resection of the lipoma
        - Reconstruction of the neural placode (pia-to-pia neurulation)
        - Expansile graft duraplasty
        - Layered closure, which may involve harvest of a myocutaneous flap or graft.
10. *Following an untethering surgery with resection of the lipoma, the child develops a CSF leak. Outline the different options available for management of this complication.*
    - Management options for a CSF leak include:
        - Cystoperitoneal shunt
        - Primary closure via suture reinforcement with or without local rotation myocutaneous flap
        - External ventricular drain placement
        - Ventriculoperitoneal shunt
        - Combination of the above (e.g., rotation flap with external drain placement)
    - Preferred options in this case remain cystoperitoneal shunting or primary closure if the CSF leak is a low-pressure leak without an associated pseudomeningocele.

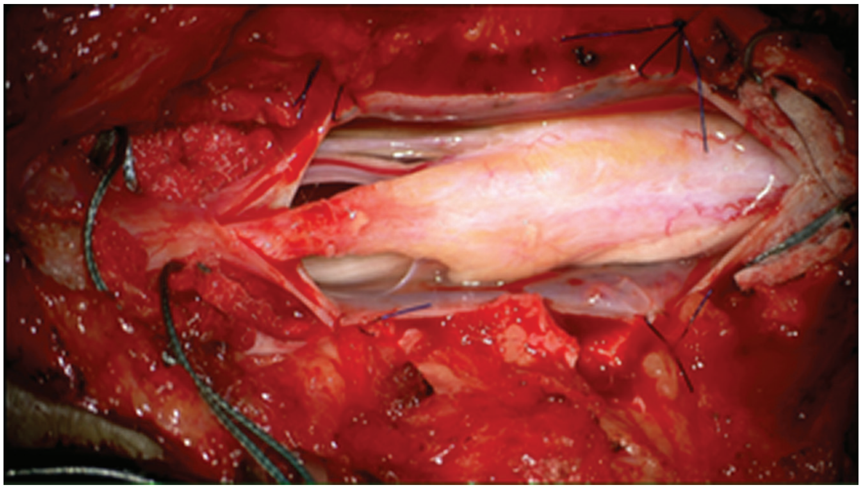

**Fig. 80.4** Intraoperative image showing exposed spinal cord lipoma after dural opening.

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# Case 81 Positional Plagiocephaly

*Han Zhuang Beh and Alexander Y. Lin*

## Clinical Presentation

- A 3-month-old boy is referred to you by his pediatrician for an abnormal head shape. It was noted by his parents at 2 months of age and they describe it as a flat spot on the right posterior portion of his head.
- The patient is otherwise healthy and meeting his developmental milestones.
- Patient's pediatrician provided the parents with positional strategies which parents stated they have been trying while at home. They have been doing so for the past 4 weeks and have not noted any improvement in head shape.

## Questions

1. What are pertinent questions to gather about the patient's history?
2. What are important anatomical landmarks to note on physical exam?
3. What are objective measures that can be done during the physical exam?
4. Differentiate plagiocephaly, brachycephaly, scaphocephaly, and trigonocephaly. What conditions cause each of these clinical findings?
5. How does posterior deformational plagiocephaly differ from unilateral lambdoid craniosynostosis?
6. If suspicious for craniosynostosis, how would you work the patient up?
7. What are the treatment options for deformational plagiocephaly?
8. When should therapy be initiated? When is it most effective?
9. What is the most common cause for treatment failure?
10. What are the complications from treatment with helmet orthotic?

## Answers

1. *What are pertinent questions to gather about the patient's history?*
   - In what position does the patient prefer to sleep (on back, on the belly, head turned in a particular direction)?
   - Does the patient's eyes completely close when he/she sleeps?
   - Has patient showed any neurological symptoms?
   - Does patient have any motor deficits or habits (torticollis)?
2. *What are important anatomical landmarks to note on physical exam?*
   - Position of the orbit, ear, cheek, and forehead in comparison to contralateral side
   - Position of the nasal tip and chin (midline or deviated to one side)
   - Level symmetry of occipital protuberances
   - Level symmetry of mastoid processes
3. *What are objective measures that can be done during the physical exam?*
   - Head circumference
   - Cranial index/cephalic index (CI; ▶**Fig. 81.1**a)
     - Cranial width/anterior to posterior (AP) length × 100
     - Mesocephalic/normocephalic: CI = 76–81 (▶**Fig. 81.1**b)
   - Cranial vault asymmetry (CVA aka difference of diagonals) and cranial vault asymmetry index (CVAI; ▶**Fig. 81.1**c)
     - Two intersecting cranial diameters drawn:
       - ○ Line from most lateral portion of frontozygomatic suture to contralateral occipital prominence
       - ○ Angle of diagonals with respect to the AP midline should be the same
     - CVA (difference of diagonals) = diagonal A − diagonal B
       - ○ 0–3 mm = normal
       - ○ No universal severity scale but insurance typically requires > 10 mm for approval of helmet orthotic
     - CVAI = ((diagonal A − diagonal B)/diagonal A) × 100
       - ○ Where diagonal A is always of greater length
       - ○ Normal < 3.5

## Answers

4. *Differentiate plagiocephaly, brachycephaly, scaphocephaly, and trigonocephaly. What conditions cause each of these clinical findings?*
   - Plagiocephaly: asymmetry due to flattening of one side of the head (▶ **Fig. 81.1**d)
     - Deformational forces
     - Unilateral lambdoid craniosynostosis (posterior plagiocephaly)
     - Unilateral coronal craniosynostosis (anterior plagiocephaly)
   - Brachycephaly: "short head" (▶ **Fig. 81.1**e)
     - CI > 81, seen in bicoronal craniosynostosis, or bilateral posterior (back-laying) deformation.
   - Scaphocephaly: "boat head" (also known as dolichocephaly, "elongated head"; ▶ **Fig. 81.1**f)
     - CI < 76, seen in sagittal craniosynostosis, or from side-laying deformation
   - Trigonocephaly: triangular forehead (▶ **Fig. 81.1**g)
     - Triangular pointed forehead with midline ridge and retruded lateral supraorbital rims bilaterally, seen in metopic craniosynostosis.
5. *How does posterior deformational plagiocephaly differ from unilateral lambdoid craniosynostosis?*
   - Both can present as posterior plagiocephaly (▶ **Fig. 81.2**)
   - Deformational plagiocephaly
     - Incidence has drastically increased after 1992, when the American Academy of Pediatrics recommended that infants be placed on their backs to sleep to prevent SIDS.
     - Parallelogram-shaped deformity
     - Ear is more anterior on flattened side
     - Frontal bone prominence ipsilateral to flattened side
     - Symmetric occipital protuberances and no distortion of cranial base
     - Appearance related problem affecting both head shape and facial symmetry
     - Studies have showed correlation between delay in motor development and deformational plagiocephaly, but no clear evidence of causative association.
   - Unilateral lambdoid craniosynostosis
     - Incidence of 3 in 100,000 live births
     - Posterior flattening occurs on side of suture fusion
     - Ear is more posterior and inferior on flattened side
     - Affected side's occipitomastoid region is enlarged and displaced inferiorly and skull base has downward slant on affected side
     - Deviation of foramen magnum toward affected side
     - Given the fact that this is a craniosynostosis, there is an increased risk for elevated intracranial pressure (ICP) and developmental delays.
6. *If suspicious for craniosynostosis, how would you work the patient up?*
   - High-resolution noncontrast CT scan of the head to evaluate cranial sutures
   - Can concomitantly evaluate ventricles and look for signs of increased ICP
   - Some centers feel that diagnosis based on physical examination alone is enough for evaluation of typical cases.
7. *What are the treatment options for deformational plagiocephaly?*
   - Goal of treatment strategies is to allow the cranium to grow preferentially in the vector that was previously affected by deformational forces; that is, treatment is nonsurgical.
   - Positioning/physical therapy
     - Strategies to limit patient's time on the affected (flat) side
       - ○ Strategic placement of stimuli (toys), repositioning crib, use of wedges/rolls to reposition patient
     - Physical therapy
       - ○ Stretching exercises (especially in patients with torticollis)
       - ○ "Tummy time"—monitored, at least 30 minutes per day
       - ○ Particularly for patients with delay in motor development
     - Evaluate after 6–8 weeks of treatment initiation. If improvement not observed move to helmet orthotic.
   - Cranial orthotic
     - May consider as primary therapy if patient has severe plagiocephaly, older patient, significant concomitant issues that affect positioning (torticollis, reflux disease), difficulty with compliance of positioning
     - Wear majority of the day (23 hours/day)
       - ○ Contraindicated in patients with hydrocephalus
8. *When should therapy be initiated? When is it most effective?*
   - Most effective during time of rapid brain/head growth
   - If possible, helmet therapy should be initiated at 4–6 months of age.
   - Efficacy of treatment starts to decrease after 6 months of age.
   - Length of therapy duration increases after 6 months of age.
9. *What is the most common cause for treatment failure?*
   - Noncompliance: both positioning and helmet therapy
10. *What are the complications from treatment with helmet orthotic?*
    - Complications from helmet are rare: dermatitis, cutaneous irritations, pressure sores
    - Failure of head shape improvement due to noncompliance with wearing helmet

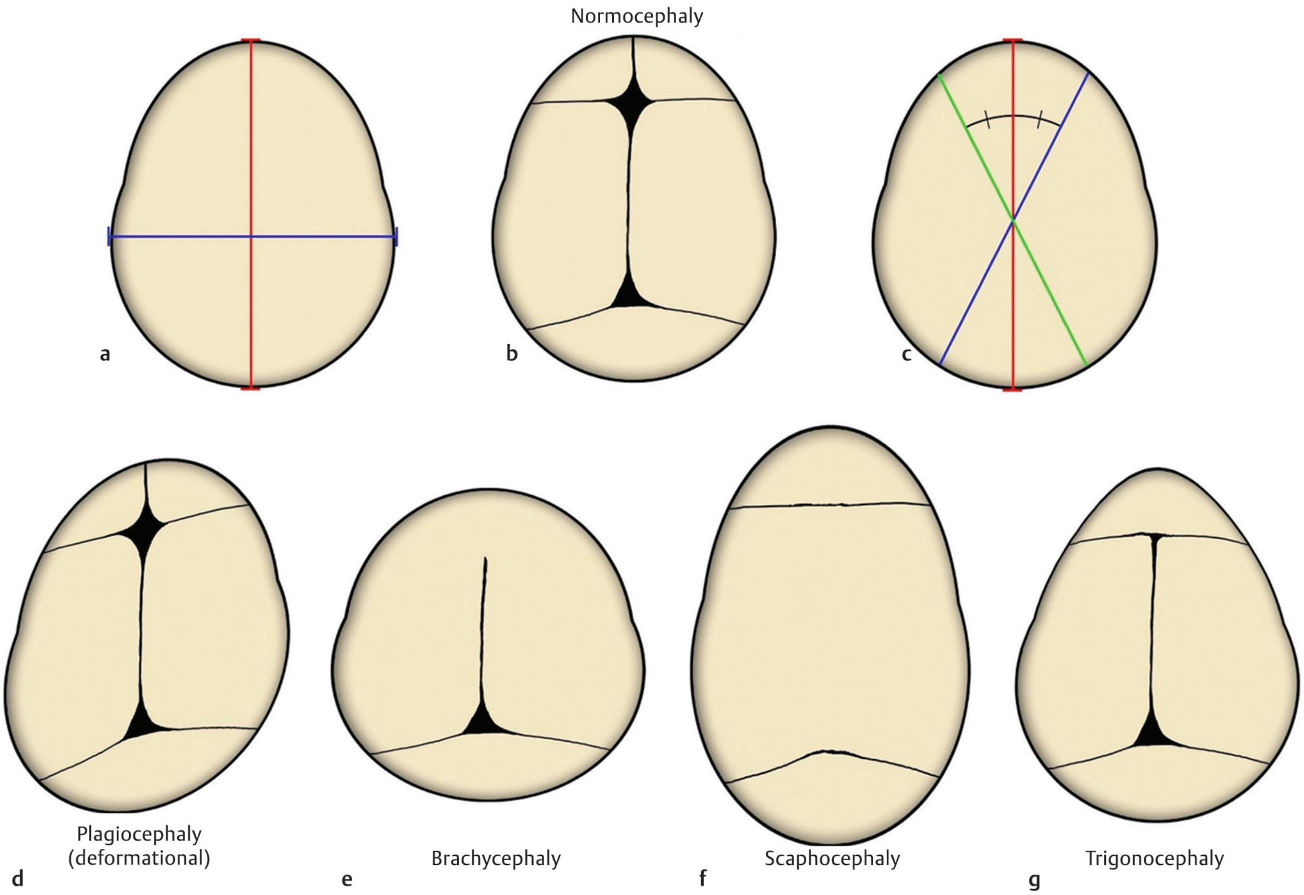
Normocephaly
a
b
c
Plagiocephaly
(deformational)
d
Brachycephaly
e
Scaphocephaly
f
Trigonocephaly
g

**Fig. 81.1** (**a**) Cephalic index, (**b**) normocephaly, (**c**) cranial vault asymmetry index, (**d**) plagiocephaly, (**e**) brachycephaly, (**f**) scaphocephaly, (**g**) trigonocephaly.

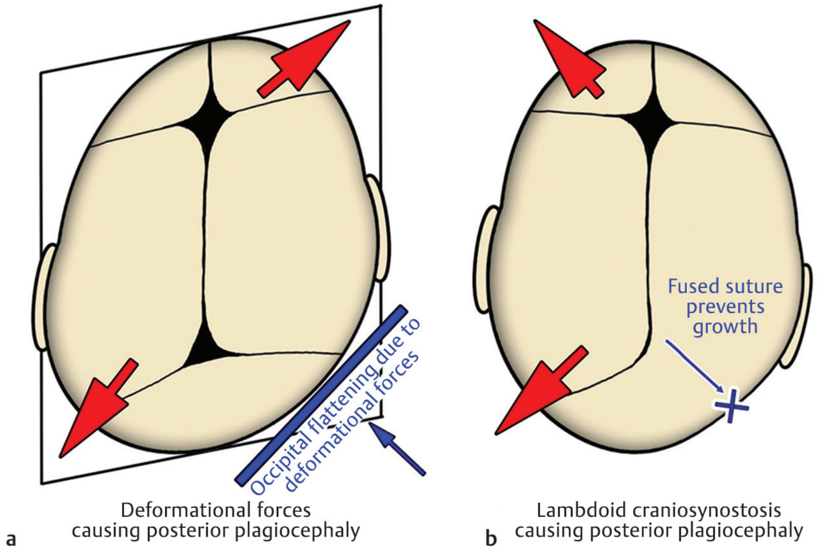
Occipital flattening due to deformational forces
Deformational forces
causing posterior plagiocephaly
a
Fused suture
prevents
growth
Lambdoid craniosynostosis
causing posterior plagiocephaly
b

**Fig. 81.2** (**a**) Positional posterior plagiocephaly. (**b**) Lambdoid craniosynostosis.

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# Case 82 Scaphocephaly—Open Repair

*Abdulrahman J. Sabbagh, Jeffrey Atkinson, Jean-Pierre Farmer, and José Luis Montes*

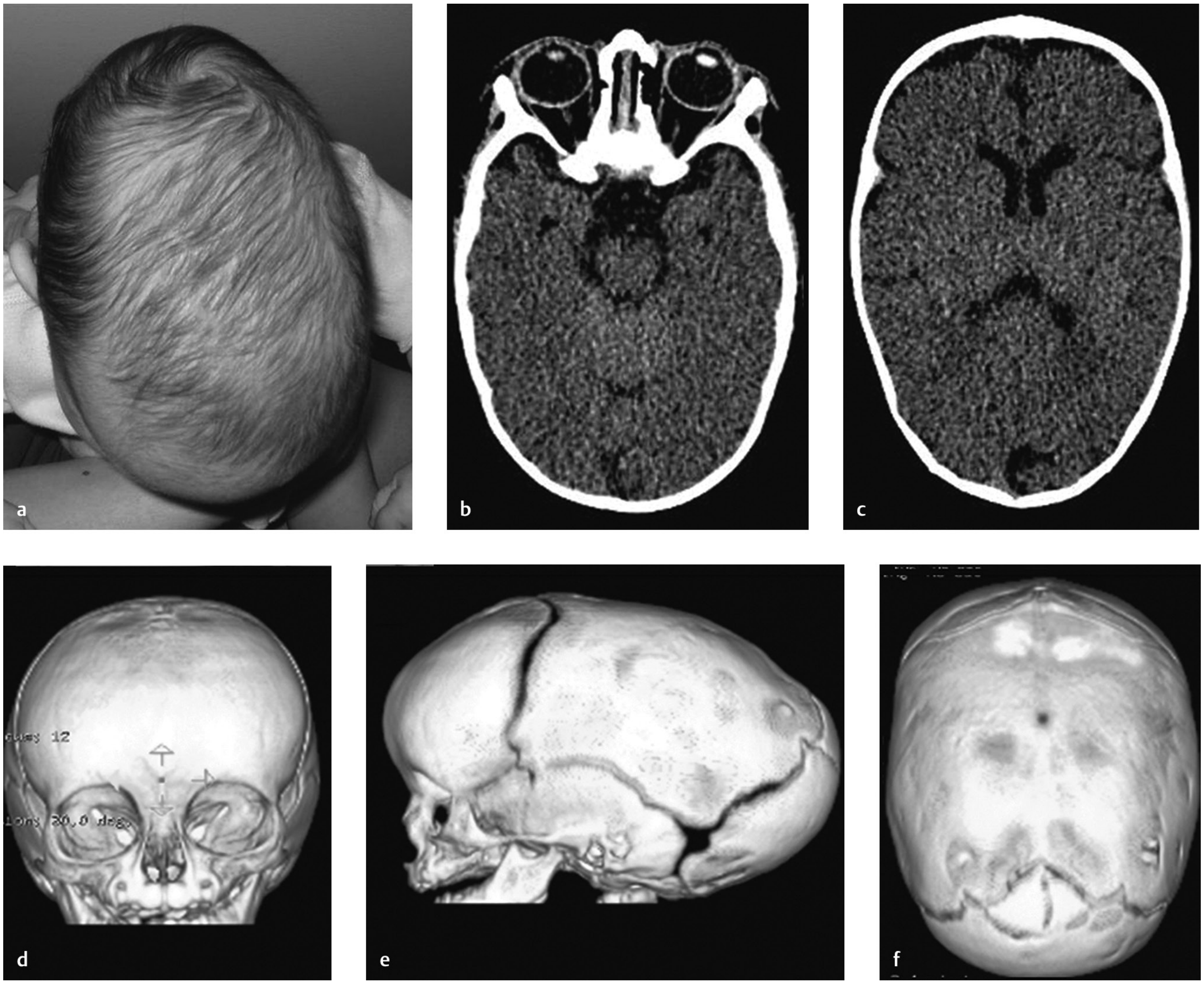

**Fig. 82.1** (**a**) Head photograph, (**b**, **c**) axial CT scan, and (**d**) three-dimensional reconstructed CT scan, (**e**, **f**) of a child with craniosynostosis.

## Clinical Presentation

- A 3-month-old child presents with an abnormal head shape since birth.
- He was born at term via a normal vaginal delivery.
- The father has a similar but more accentuated head shape.
- The physical examination is otherwise normal.
- A CT scan of the head is done and shown in ▶**Fig. 82.1**.

## Questions

1. Describe the head shape and the CT scan. What is the diagnosis?
2. What is scaphocephaly?
3. How common is scaphocephaly?
4. What is the prognosis of scaphocephaly?
5. What is the risk of developing hydrocephalus in patients diagnosed with craniosynostosis? What is the risk for this patient?
6. What are some treatment options and their possible related morbidities?
7. What is the accepted hypothesis for the pathogenesis of deformities caused by craniosynostosis?
8. What is more clinically important, timing of head and brain growth or timing of normal sutural closure, and why?

## Answers

1. *Describe the head shape and the CT scan. What is the diagnosis?*
   - The photograph, plain CT, and three-dimensional reconstructed CT scans show an elongated boat-like skull with a closed sagittal suture (▶ **Fig. 82.1**).
   - Also seen is frontal and occipital bossing.
   - The diagnosis is scaphocephaly.
2. *What is scaphocephaly?*
   - Scaphocephaly can be described as a boat-shaped head, caused by synostosis of the sagittal suture, leading to a long and thin skull.[1]
   - There is bifrontal and occipital symmetric compensatory bossing.
   - There is usually a midline ridge, and there may be a saddle deformity where the synostosis began.
   - It may be evident at birth and progresses to become more pronounced with time.
   - It is the most common type of synostosis and is more common in males than females.
3. *How common is scaphocephaly?*
   - Epidemiology of scaphocephaly is described below.
     - Incidence is 2 to 10 per 10,000 live births.[2]
     - It comprises 55 to 64% of synostosis surgical cases (most common type).
     - Seventy to 85% of patients are male.[1]
     - Six to 10% of cases are familial.
     - It may follow an autosomal dominant inheritance pattern.[2]
     - There is 38% penetrance.[2]
4. *What is the prognosis of scaphocephaly?*
   - This deformity, like other forms of craniosynostosis, usually progresses with time and will have an important impact on the growing child's social and psychological status.[3]
   - These children usually would develop normally from a neurologic standpoint.
   - There is a slight risk of increased intracranial pressure (ICP).[3]
5. *What is the risk of developing hydrocephalus in patients diagnosed with craniosynostosis? What is the risk for this patient?*
   - Renier et al in 1982 measured ICP using epidural sensor in 92 patients for 12 to 24 hours.[4] He found the following results as they relate to ICP.
     - In one-suture synostosis, 62% were normal, 24% were borderline, and 14% were high.
     - In several-suture synostosis, 19% were normal, 34% were borderline, and 47% were high.
   - The incidence of hydrocephalus in sagittal craniosynostosis is remarkably rare (~0.3%), except in patients with prematurity or those who have been shunted.[5]
   - On the other hand, in syndromic craniosynostosis (Crouzon syndrome, Pfeiffer syndrome, Apert syndrome, etc.), the incidence of hydrocephalus may range from 20 to 40%.[5,6,7] The former two are more likely to be associated with shunt dependent hydrocephalus than the latter.[7]
6. *What are some treatment options and their possible related morbidities?*
   - Ideally, surgical repair should be done at 2 to 3 months of age. When done before 3 months of age, the following options are available:
     - Midline strip craniectomy[8]
       - A 4- to 8-cm wide craniectomy is performed, from the coronal to just posterior to the lambdoid sutures.
       - Bilateral barrel stave osteotomies are then done (▶ **Fig. 82.2**).
       - Blood transfusion is common.
       - There is up to a 10% chance of having a residual bone defects.
       - Good cosmetic results can be obtained (▶ **Fig. 82.3**).
     - Pi procedure[9,10]
       - This procedure immediately provides anteroposterior shortening of the skull.
       - It may also give a better cosmetic result.
     - Endoscopic strip with molding helmet therapy
       - This method significantly lowers the rate of transfusion.[11]
   - In cases where treatment is contemplated late (after 6 months of age), the following options are available:
     - Variations of the Pi procedure[9]
     - Total cranial vault reconstruction[12]

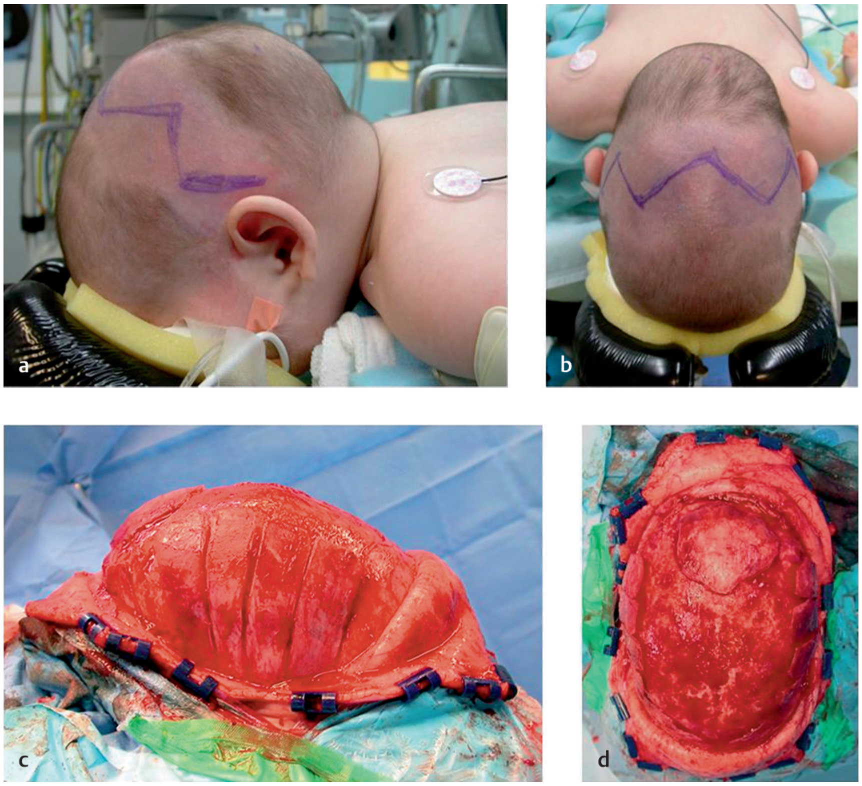

**Fig. 82.2** Intraoperative photograph showing (**a, b**) incision location as well as (**c, d**) strip craniectomy with bilateral barrel stave osteotomies.

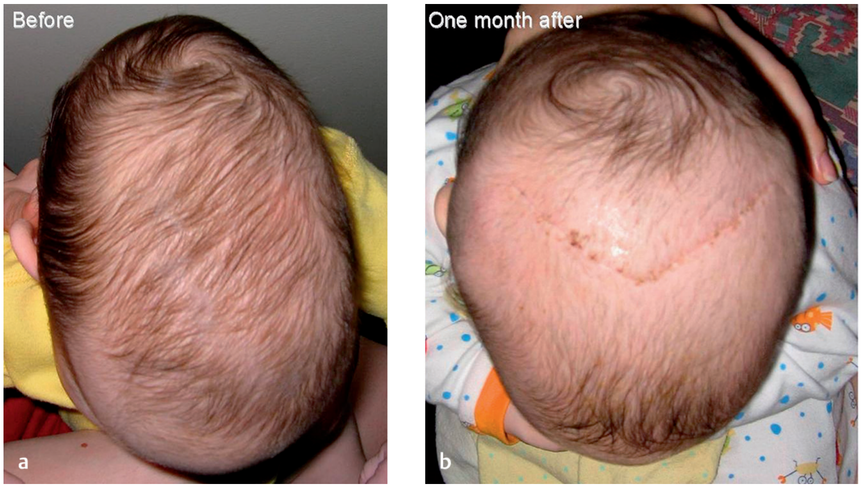

**Fig. 82.3** (**a**) Preoperative and (**b**) postoperative photographs showing comparison and improvement in cosmetic result after strip craniectomy with barrel stave osteotomies shown in **Fig. 82.2**.

## Answers (*continued*)

7. *What is the accepted hypothesis for the pathogenesis of deformities caused by craniosynostosis?*
   - An older hypothesis described by Virchow (in 1851) states, “Craniosynostosis is caused by a lack of growth perpendicular to the fused suture and compensatory growth parallel to the suture in the calvarial vault.”[13]
   - The currently accepted hypothesis devised by Delashaw et al states, “The calvarial bones directly adjacent to a fused suture act as a single bone plate with decreased growth potential.”[13]
   - Asymmetrical bone deposition occurs at the sutures along the perimeter of the bone plate with increased bone deposition at the outer margin.[13]
   - Nonperimeter sutures in line with the fused suture deposit bone symmetrically at their sutural edges.[13]
   - Perimeter sutures adjacent to the fused suture compensate to a greater degree than other distant sutures.[13]
8. *What is more clinically important, timing of head and brain growth or timing of normal sutural closure, and why?*
   - Timing of head and brain growth is more clinically significant than normal suture closure.[1]
     - The brain doubles in size by 6 months of age and doubles again by 2 years of age.
     - The skull is 35% of its adult size at birth, and 90% of the adult size is reached by age 7 years.

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# Case 83 Scaphocephaly—Endoscopic Repair

*Ananth K. Vellimana, Kamlesh B. Patel, and Matthew D. Smyth*

## Clinical Presentation

- A 2-month-old boy was referred to clinic for evaluation of abnormal head shape which was noted at birth.
- He was born at full term gestation. No other abnormalities were identified on physical exam.
- He does not have any relatives with similar problems.
- Examination and imaging findings are shown in ▶ **Fig. 83.1**.

## Questions

1. What is the clinical diagnosis?
2. Describe scaphocephaly.
3. What is the cephalic index?
4. What are the different treatment options for sagittal synostosis?
5. What are the advantages of the endoscopic approach?
6. What are the indications for endoscopic repair?
7. Describe the endoscopic technique for repair of scaphocephaly.
8. What is the expected postoperative course of patients undergoing endoscopic repair?
9. What are the common complications associated with the endoscopic approach as compared to the open approach?

## Answers

1. *What is the clinical diagnosis?*
   - Images demonstrate an infant with elongated head shape along with biparietal narrowing, and frontal and occipital bossing (▶ **Fig. 83.1**a–c).
   - The sagittal suture appears to be closed (▶ **Fig. 83.1**d–i). This suggests a diagnosis of scaphocephaly secondary to sagittal synostosis.
2. *Describe scaphocephaly*.
   - Scaphocephaly is a skull deformity wherein the head appears narrow and elongated, resembling an inverted boat (▶ **Fig. 83.1**b).
   - It occurs due to sagittal synostosis, which is the most common form of isolated craniosynostosis.[1]
   - Normally, fusion of the sagittal suture occurs between the third and fourth year of life. In utero closure of the suture leads to skull growth in an anteroposterior direction, leading to scaphocephaly.
   - The degree of suture fusion and severity of skull deformity can markedly vary among individuals.
   - A ridge can often be palpated along the fused sagittal suture. Frontal bossing, occipital bossing, and coronal constriction are some other features that may be seen.
3. *What is the cephalic index?*
   - Cephalic index (also known as cranial index) is the ratio of maximum width of the skull (distance from euryon to euryon) to maximum length of the skull (distance from glabella to opisthocranion).[2]
   - It can be measured directly with calipers or indirectly using 3D photography or CT scans.
   - Cephalic index is a useful tool for pre- and postoperative assessment of skull deformity. It is considered normal between 75 and 83%. Patients with scaphocephaly have decreased cephalic index.
4. *What are the different treatment options for sagittal synostosis?*
   - Surgical correction is the mainstay of treatment for sagittal synostosis. The principal goals of surgery are:
     - Improvement of aesthetic appearance of the skull
     - Relief of any constriction of the brain, thereby preventing the development of increased intracranial pressure
   - The available surgical options depend on age of the child; these include:
     - For children less than 6 months of age, both endoscopic strip craniectomy with postoperative helmet molding therapy and open calvarial vault reconstruction are good surgical options.[1,3]
       - ○ Ideally, endoscopic repair should be performed within the first 4 months of age, since strip craniectomies may be less efficacious in children older than 4 months.[1,4,5]
       - ○ Open procedures often require blood transfusion and are usually performed at or after 3 months of age to avoid the nadir of physiological anemia of infancy.

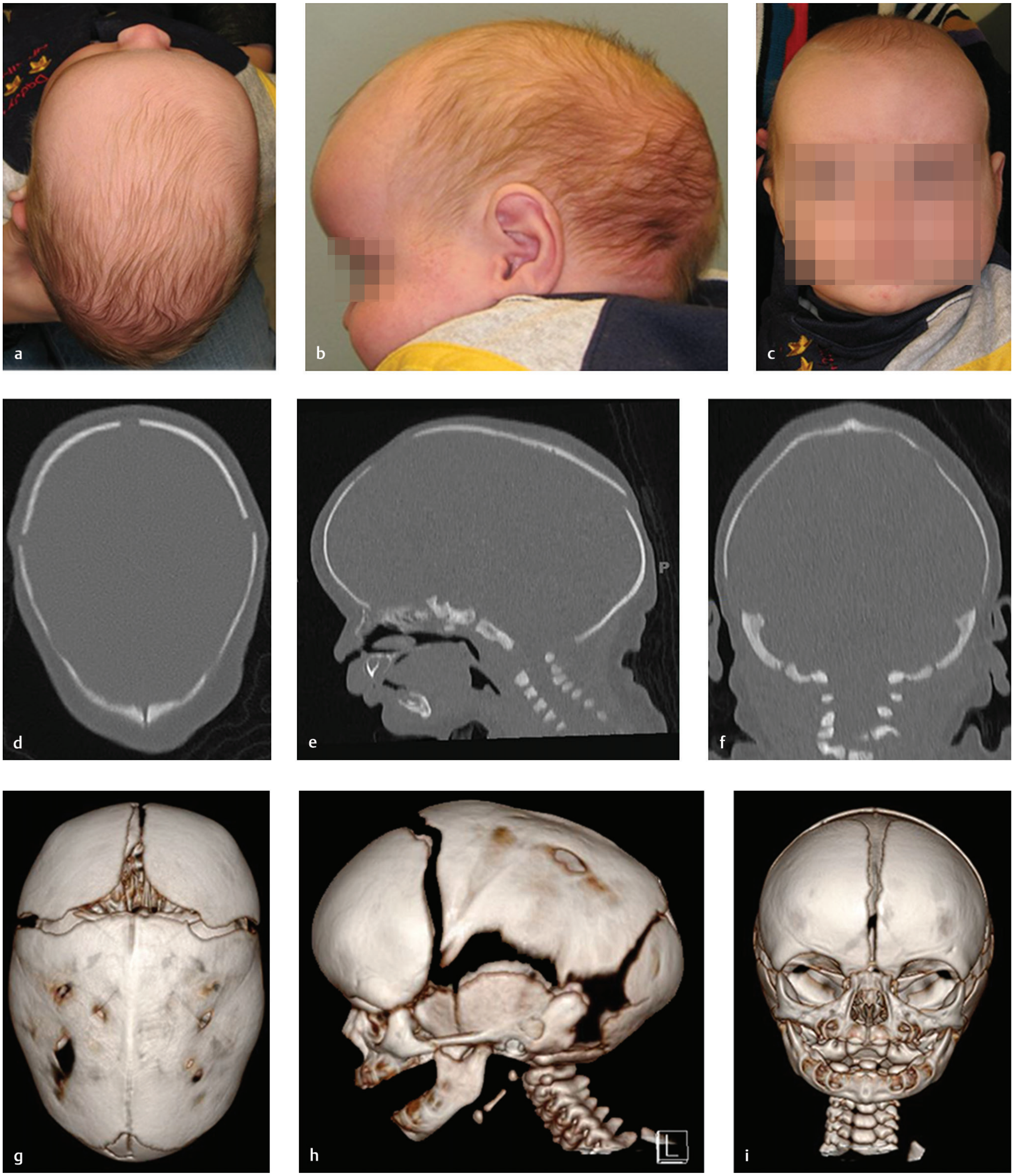

**Fig. 83.1** Infant with sagittal synostosis: Bird's eye (**a**), lateral (**b**), and anteroposterior (**c**) views on physical exam; axial (**d**), sagittal (**e**), coronal (**f**) CT scan images; Bird's eye (**g**), lateral (**h**), anteroposterior (**i**) views of three-dimensional CT scan reconstructions.

## Answers (*continued*)

  - For children older than 6 months of age, open reconstruction is the only viable surgical option.
    - The open techniques utilized at the authors' institution include subtotal calvarial vault reconstruction or a modified Pi procedure.[6] The subtotal calvarial vault reconstruction procedure is used in older patients in whom the cranial bone is less malleable. This involves removal and reshaping of the parietal bones. Additional osteotomies are performed in the frontal and occipital bones to accommodate parietal expansion. The modified Pi procedure utilized at the authors' institution involves frontal, bilateral parietal, and occipital barrel stave osteotomies and outfractures to accommodate parietal expansion. In addition, the midsagittal strip is shortened to the frontal bone.[1,6,7]

5. *What are the advantages of the endoscopic approach?*
   - Advantages of the endoscopic approach include:
     - Significantly shorter operative time
     - Decreased blood loss
     - Decreased transfusion requirement
     - Shorter length of hospital stay[1]
   - The improvement obtained in cephalic index is comparable between open and endoscopic procedures.[1,3,6] Both open and endoscopic procedures result in similar cranial vault volumes 1 year postoperatively for sagittal synostosis, and these volumes are comparable to control subjects.[6]
   - Endoscopic approaches also have a decreased cost and are less of a burden on health care resources.[8]
6. *What are the indications for endoscopic repair?*
   - Age < 6 months
   - Commitment to comply with helmet therapy
7. *Describe the endoscopic technique for repair of scaphocephaly.*
   - The children are administered general endotracheal anesthesia.
   - Two peripheral IV lines are placed. The authors routinely do not use an arterial line or Foley catheter for endoscopic craniosynostosis procedures.
   - The team includes a pediatric neurosurgeon and a pediatric craniofacial plastic surgeon.
   - Patients are positioned in the modified "sphinx" position with the head extended and resting on a Doro head holder (see ▶ **Fig. 83.2**g).
   - An extension test is performed by the anesthesiologist prior to positioning to make sure that the endotracheal tube remains optimally positioned in the extended sphinx position.
   - Two incisions are made—one is slightly posterior to the anterior fontanelle and the other incision is slightly anterior to the lambda (▶ **Fig. 83.2**g).
   - After subgaleal dissection, burr holes are placed at both incisions.
   - The dura is stripped from the overlying bone and the burr holes are expanded across the midline using a combination of curettes and Kerrison rongeurs.
   - At the anterior site, the osteotomy is connected to the anterior fontanelle using bone scissors. This provides an access site for the endoscope.
   - Under endoscopic visualization, the dura is stripped from the overlying bone (▶ **Fig. 83.2**h).
   - The authors' prior technique involved a wide vertex craniectomy and bilateral temporal and parietal wedge osteotomies.[6,9] At present, the authors perform a narrow (~2.5 cm wide) craniectomy of the fused sagittal suture using bone scissors (▶ **Fig. 83.2**i, j).[1,6,7,10,11]
   - Of note, in the endoscopic technique, there is no modification of the frontal or occipital bones.
   - Under endoscopic visualization, hemostasis is then obtained at the bone edges utilizing a suction/Bovie electrocautery set at 50W (▶ **Fig. 83.2**k).
   - After irrigation and additional hemostasis, the wound is closed in layers with absorbable stitches, and a surgical skin adhesive for the skin edges.
   - The patient is then extubated and transferred to the post anesthesia care unit, followed by admission directly to the regular surgical floor.
8. *What is the expected postoperative course of patients undergoing endoscopic repair?*
   - Patients undergoing endoscopic repair are typically admitted to the neurosurgical floor for observation.
   - Hemoglobin and hematocrit studies are sent approximately 4 to 6 hours postoperatively to allow some time for equilibration of intraoperative intravenous fluid administration. A hematocrit threshold of < 18% is used for blood transfusion, and the necessity for transfusion after an endoscopic repair is rare.
   - Patients are typically discharged home on postoperative day one. They are fitted with a cranial molding helmet in the first postoperative week, and are expected to wear the helmet for 23 hours a day. They are followed for compliance regularly, until completion of helmet therapy at approximately 1 year of age. Two or three helmets are typically utilized during this period.
9. *What are the common complications associated with the endoscopic approach as compared to the open approach?*
   - Common complications associated with endoscopic repair include[7,12,13,14,15]:
     - Dural tears (4%)
     - Postoperative infections (1%)
     - Blood loss requiring transfusion (up to 5–6%)
     - Conversion to open procedures (less than 1%)

## Answers (*continued*)

  - Calvarial defect (less than 1%)
  - Venous air embolism (less than 1%)
- Other complications, also reported with the open approach on occasion, include the ones listed below:
  - Sagittal sinus lesion
  - Iodine-induced blisters
  - Pulmonary complications
- While these complications may be technique and center dependent, they provide the reader a general idea of what to expect when comparing the two approaches.

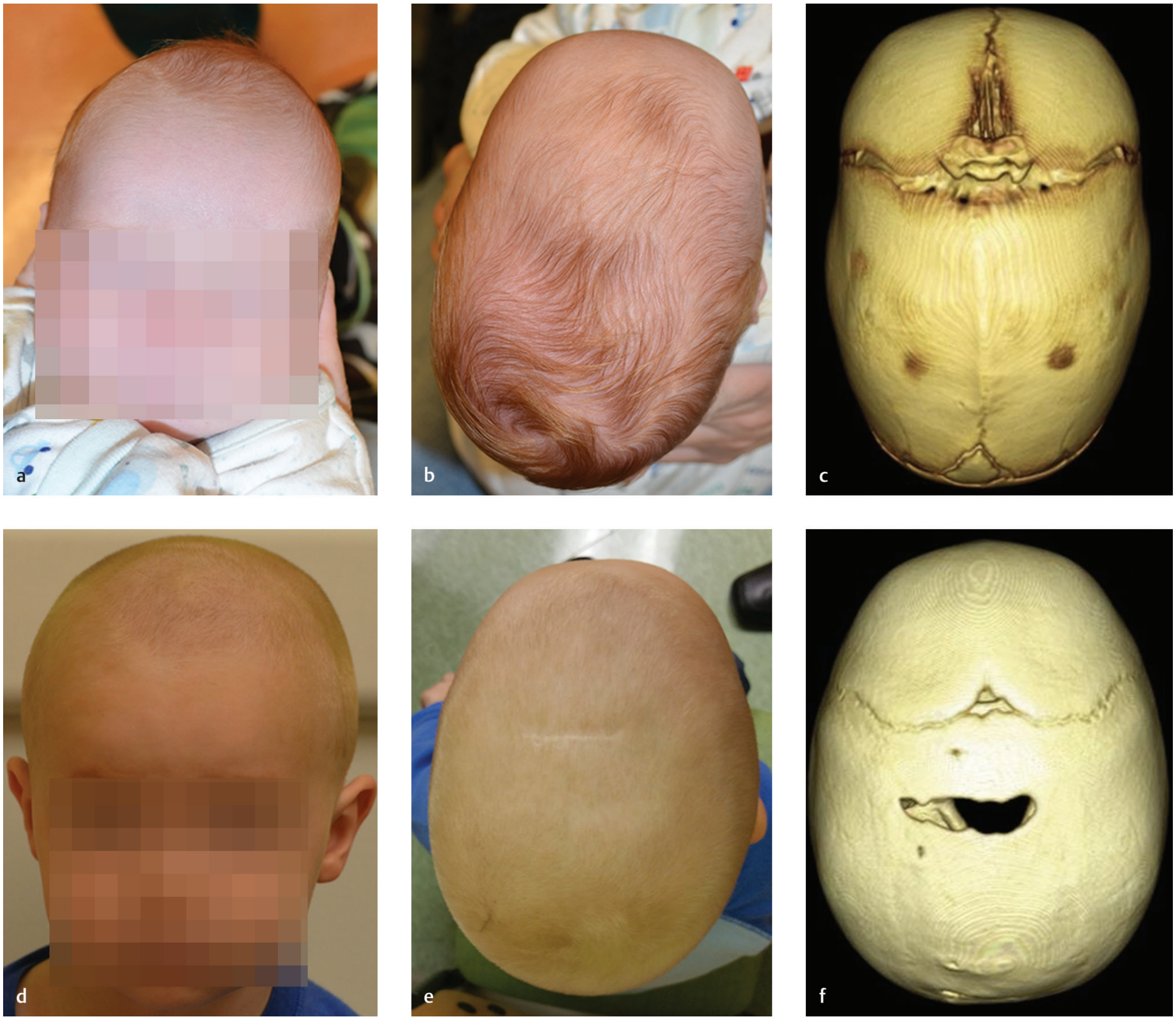

**Fig. 83.2** Preoperative (**a**, **b**, **c**) and 2-year postoperative (**d**, **e**, **f**) anteroposterior (**a**, **d**) and bird's eye view images (**b**, **e**), and bird's eye view three-dimensionally reconstructed CT scan images (**c**, **f**) of a child with sagittal synostosis who underwent endoscopic strip suturectomy and postoperative helmet molding therapy.

(*Continued*)

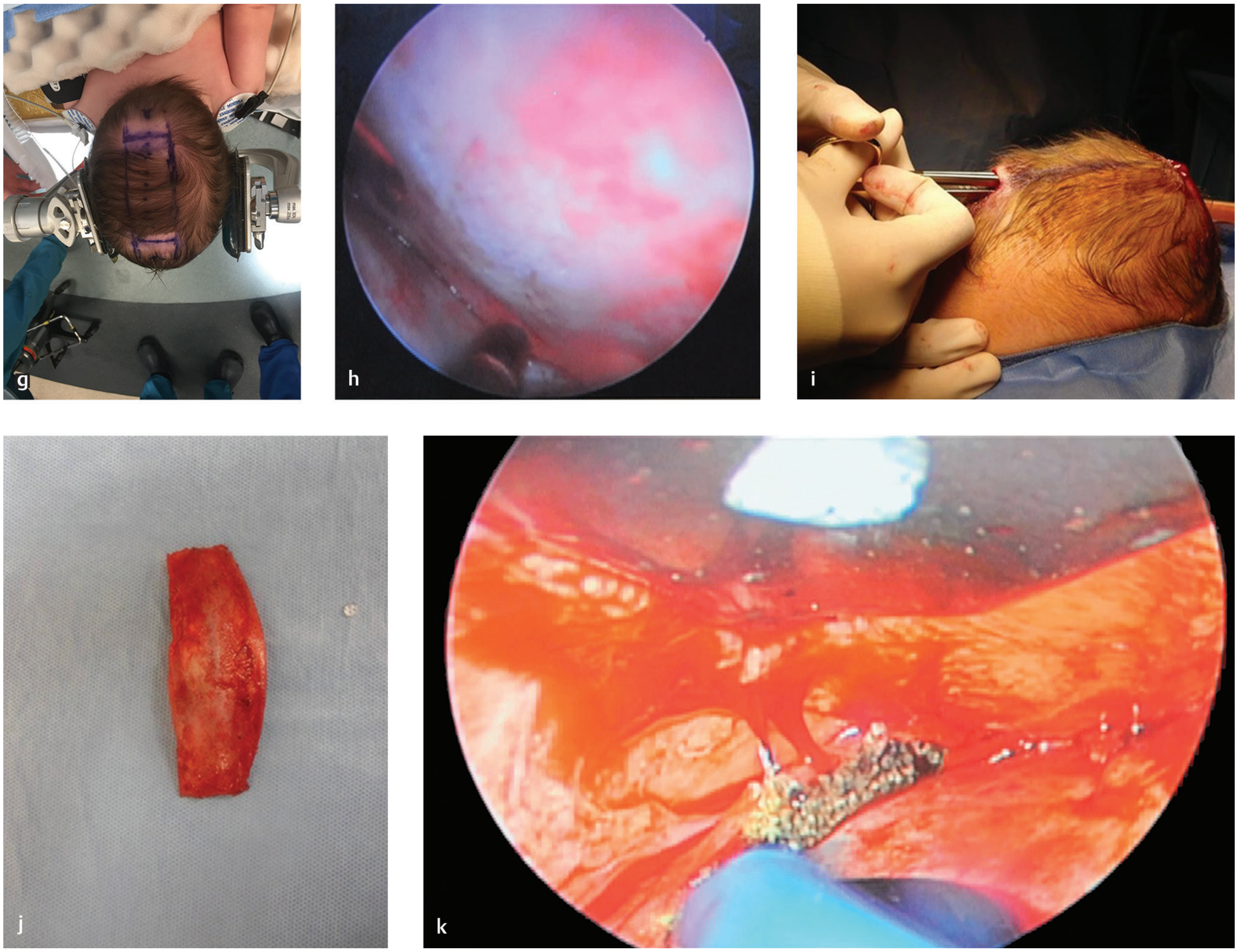

**Fig. 83.2 (*continued*)** (**g**) The patient is positioned in a "sphinx" position on the Doro head holder. (**h**) Under endoscopic visualization, dura is stripped from the overlying bone/suture. (**i, j**) Approximately 2.5 cm vertex suturectomy is performed with bone scissors. (**k**) Bone edges are coagulated under endoscopic visualization using a suction/Bovie electrocautery set at 50W. J&K Scalp-Dura Retractor is used to protect the surrounding tissue during cauterization.

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# Section V Intracranial Pathology: Functional Neurosurgery

# Case 84 Tic Douloureux

*Burak Sade and Joung H. Lee*

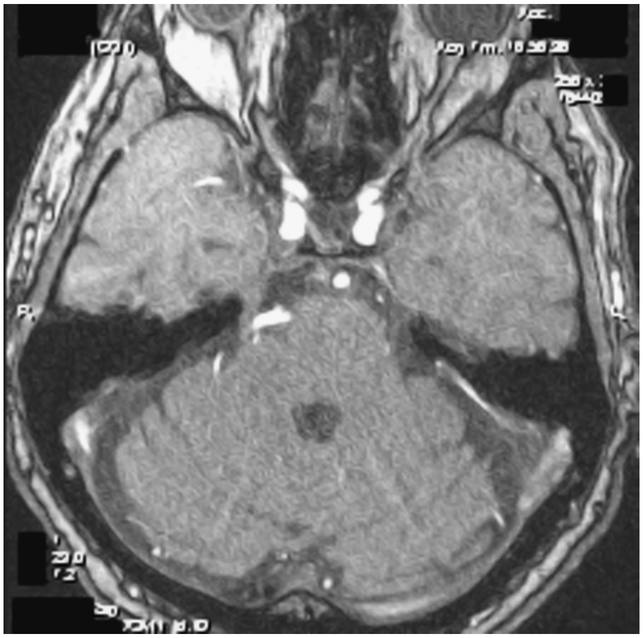

**Fig. 84.1** MRI scan of the brain. Three-dimensional axial image of the circle of Willis demonstrating a prominent vascular loop ventral to the trigeminal nerve root entry zone on the right side.

## Clinical Presentation

- An 83-year-old right-handed man presents with a 2-year history of neuralgia-like pain on the right side of his face involving the ophthalmic (V1) and maxillary (V2) divisions of the trigeminal nerve.
- He has been taking carbamazepine and topiramate, which provided minimal relief, and says he no longer wishes to use these medications.
- His neurologic evaluation is within normal limits.
- MRI of the brain is shown in ▶**Fig. 84.1**.
- The patient underwent a microvascular decompression (MVD) procedure. At the time of surgery, the trigeminal nerve was found to be in severe compression by a redundant vertebrobasilar complex ventrally and inferiorly, and by the anterior inferior cerebellar artery superiorly. Decompression was achieved using small pieces of Teflon paddies.
- He experienced immediate and complete recovery of his pain following surgery.

## Questions

1. What are the different types of facial pain syndromes?
2. What are the characteristics of the pain in trigeminal neuralgia?
3. What are the main nuclei of the trigeminal nerve, and what functions do they serve?
4. What is the initial management of a patient with trigeminal neuralgia?
5. What are the surgical indications?
6. What are the other treatment options?
7. Which vessel is the most common culprit of the compression?
8. What are the efficacies of MVD and other treatment options in trigeminal neuralgia?

## Answers

1. *What are the different types of facial pain syndromes?*
   - According to the classification proposed by Burchiel[1]:
     – Trigeminal neuralgia (type 1—predominant episodic and type 2—predominant constant) or symptomatic trigeminal neuralgia (in multiple sclerosis)
     – Trigeminal neuropathic pain
     – Trigeminal deafferentation pain
     – Postherpetic neuralgia
     – Atypical facial pain
2. *What are the characteristics of the pain in trigeminal neuralgia?*
   - Characteristics of pain in trigeminal neuralgia[1]:
     – Sharp, shooting, electric shock-like pain
     – Momentary or lasting only a few seconds
     – Very intense
     – Common provoking factors include touching, washing the face, teeth brushing, make-up, chewing, talking, eating, and cold breeze.
     – May involve one or more branches of the trigeminal nerve
3. *What are the main nuclei of the trigeminal nerve, and what functions do they serve?*
   - The principal sensory or main nucleus: located in the upper pons, it conveys tactile and pressure senses from the face.
   - The mesencephalic nucleus: located near the central gray matter of the upper fourth ventricle, it conveys pressure and kinesthetic senses from the teeth, hard palate, and jaw.
   - The spinal trigeminal tract and nucleus: extends from the upper cervical spine to the mid pons, it is divided into three parts (pars caudalis, pars interpolaris, and pars oralis), which convey sensation of pain and temperature from different parts of the face (▶**Fig. 84.2**).
   - The motor nucleus relays fibers to the muscles of mastication and plays part in the jaw jerk reflex.
   - The ventral and dorsal trigeminothalamic tracts relay sensory information to the ventroposterior medial nucleus of the thalamus.[2]
4. *What is the initial management of a patient with trigeminal neuralgia?*
   - Initial management in trigeminal neuralgia is medical.
   - Medications that are most commonly used include carbamazepine, gabapentin, lamotrigine, and Trileptal (Novartis, East Hanover, NJ).
   - Among these, carbamazepine is the most widely used.
   - In addition, antidepressants or narcotic analgesics and steroids during severe pain episodes may provide temporary relief.
5. *What are the surgical indications?*
   - Indication for surgical treatment include failure of medical therapy, intolerance to the medications, and patients who do not like to take medications for a long time.
6. *What are the other treatment options?*
   - Other treatment options include[3,4,5]:
     – MVD
     – Percutaneous techniques: glycerol rhizotomy (GR), balloon compression (BC), and radiofrequency rhizotomy (RF)
     – Gamma knife radiosurgery (GKRS)

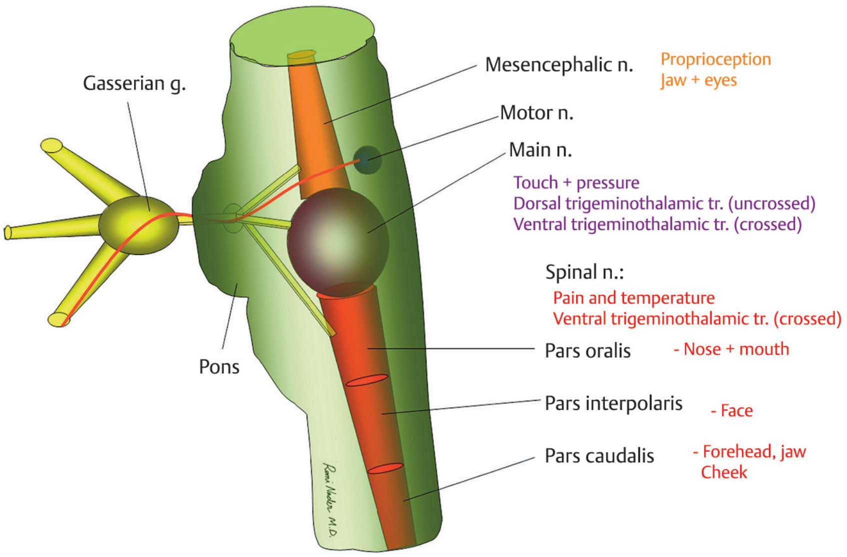
Gasserian g.
Mesencephalic n.
Proprioception
Jaw + eyes
Motor n.
Main n.
Touch + pressure
Dorsal trigeminothalamic tr. (uncrossed)
Ventral trigeminothalamic tr. (crossed)
Spinal n.:
Pain and temperature
Ventral trigeminothalamic tr. (crossed)
Pars oralis
- Nose + mouth
Pars interpolaris
- Face
Pars caudalis
- Forehead, jaw
Cheek
Pons

**Fig. 84.2** Artist's rendering of the nuclei of the trigeminal nerve, their locations in the brainstem, their functions, and associated tracts. g, ganglion; n, nucleus; tr, tract.

## Answers (*continued*)

7. *Which vessel is the most common culprit of the compression?*
   - Superior cerebellar artery (75%)[3]
8. *What are the efficacies of MVD and other treatment options in trigeminal neuralgia?*
   - In a review by Taha and Tew,[4] the initial pain relief was 98% in MVD and RF, 93% in BC, and 91% in GR.
   - Ten years after surgery, excellent results were seen in 70% of cases who underwent MVD.[3]
   - Pain recurrence was seen in 15% cases of MVD, 21% in BC, 23% in RF, and 54% in GR.[4]
   - Postoperative facial numbness and corneal anesthesia incidences were 2% and none in MVD, 60 and 4% in GR, 72 and 2% in BC, 98 and 7% in RF, respectively.[4]
   - Therefore, when V1 or multiple divisions including V1 are involved, MVD is the preferred option to minimize the risk of corneal anesthesia.
   - GKRS has been reported to be effective in 50 to 80% of the patients, with ~25% pain recurrence.[5]
     – The risk of facial numbness and dysesthesias increase with higher radiation dose in this technique.

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# Case 85 Hemifacial Spasm

*Bassem Yousef Sheikh*

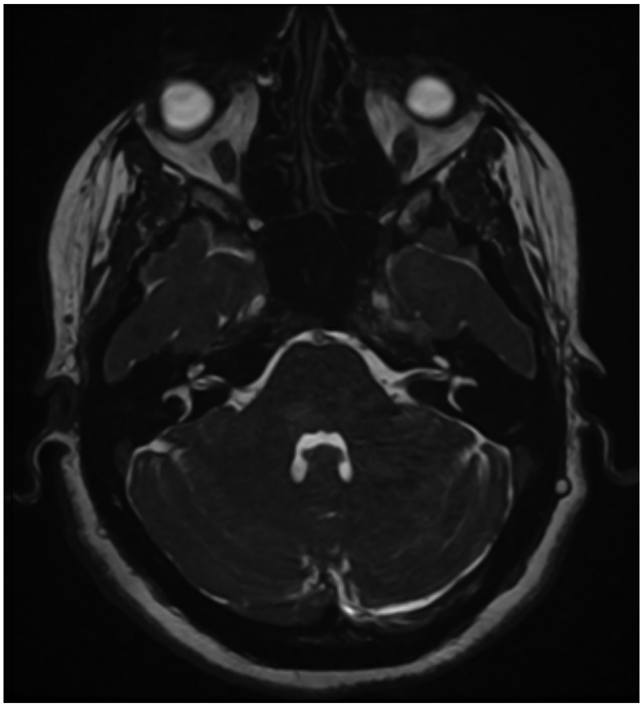

**Fig. 85.1** T2-weighted MR image at the level of the posterior fossa.

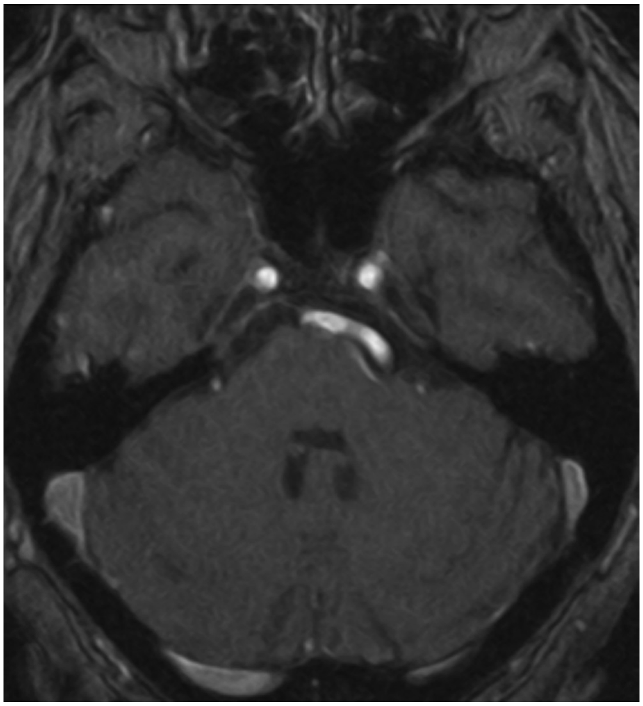

**Fig. 85.2** T2-weighted MR image of another patient with HFS at the level of the posterior fossa.

## Clinical Presentation

- A 51-year-old woman presented with a history of 13 years of spasmodic facial twitching on the left side.
- The twitching involves the left eye lid leading to intermittent spasmodic closure of the eye.
- These attacks are exacerbated with stress, and are sometimes associated with blurred vision.
- Clinical examination did not reveal any neurologic deficit.

## Questions

1. What is the most likely diagnosis?
2. Describe the clinical types of hemifacial spasm (HFS).
3. Give possible differential diagnoses.
4. Describe the pathophysiological bases of HFS.
5. How will you investigate this patient?
6. Interpret the MRI images (▶**Fig. 85.1**, ▶**Fig. 85.2**, ▶**Fig. 85.3**, and ▶**Fig. 85.4**).
7. What is the underlying etiology of HFS?
8. What are the surgical and nonsurgical therapeutic options?
9. Briefly describe your surgical procedure for HFS.
10. What is the expected surgical outcome?
11. What are the possible surgical complications?

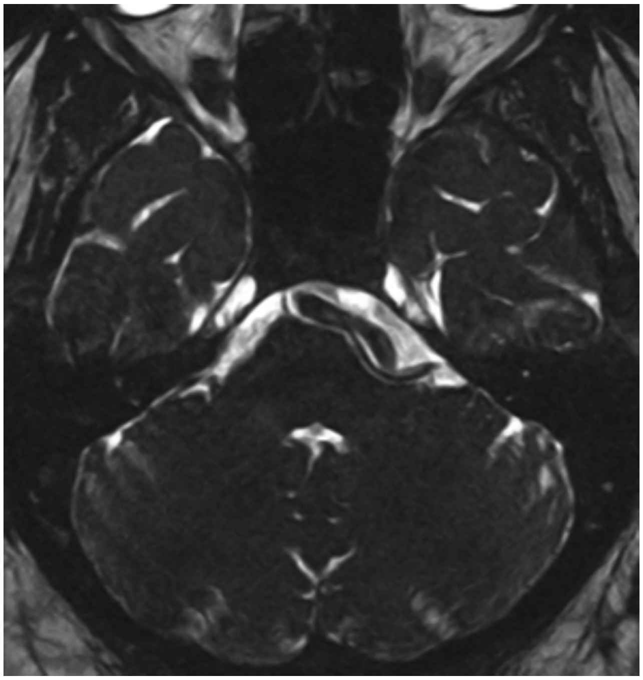

**Fig. 85.3** T1- and 4 T2-weighted MR images at the level of the posterior fossa and cerebellopontine angle (C). Vertebral digital subtraction angiography of the same patient.

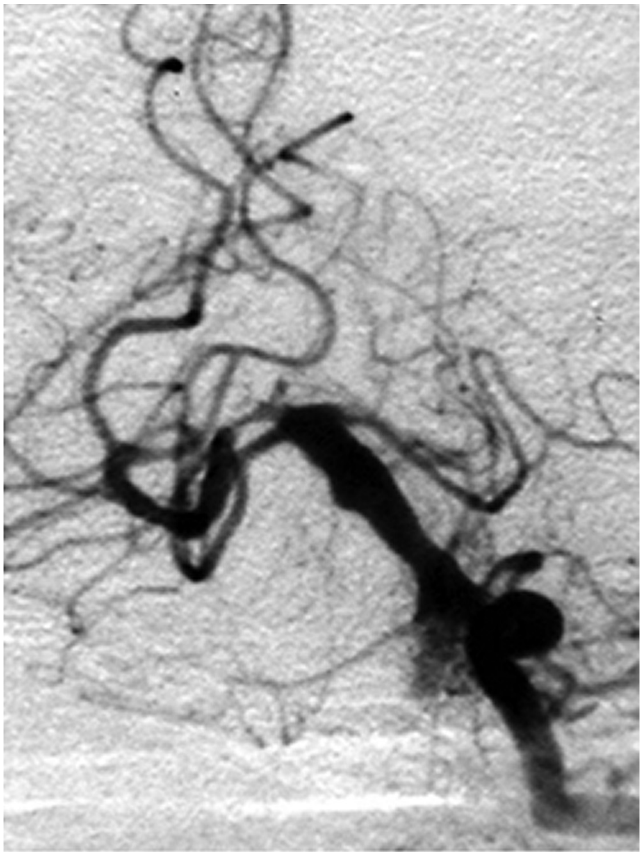

**Fig. 85.4** Vertebral artery digital subtraction angiogram.

## Answers

1. *What is the most likely diagnosis?*
   - The most likely diagnosis is hemifacial spasm (HFS).
2. *Describe the clinical types of hemifacial spasm (HFS).*
   Two types are recognized:
   - Typical HFS: symptoms begin within the orbicularis oculi muscles and progress caudally.
   - Atypical HFS: symptoms begin within the buccal muscles and progress rostrally.
3. *Give possible differential diagnoses.*
   Differential diagnoses include:
   - Benign essential blepharospasm
   - Craniofacial tremor
   - Facial chorea
   - Tics
   - Facial myokymia
   - Hemifacial twitching secondary to mass compression.
4. *Describe the pathophysiological bases of HFS.*
   Pathophysiology of HFS:
   - HFS is a neuromuscular disorder that is characterized by paroxysmal bursts of involuntary, intermittent or continuous clonic movements that progress to sustained tonic activity occurring in the muscles innervated by the facial nerve.
   - It is believed to be secondary to irritation of the facial nerve nucleus leading to its hyperexcitability.
   - It may also be due to irritation of the proximal facial nerve segment, which may cause ephaptic transmission within the nerve.
   - Either mechanisms explain the rhythmic involuntary myoclonic contractions observed in HFS.
   - The disorder presents almost always unilaterally, although bilateral involvement may occur rarely in severe cases.
   - Typically, HFS results secondary to vascular cross-compression of the myelinated facial nerve at or proximal to the junctional area of central and peripheral myelin (the root exit zone) of the nerve.
5. *How will you investigate this patient?*
   Investigation of this patient includes the following:
   - Neurophysiologic testing:
     - Spread and variable synkinesis on blink reflex testing and high-frequency discharges on

## Answers (*continued*)

electromyography (EMG; with appropriate clinical findings) are diagnostic.

    - ○ Stimulation of one branch of the facial nerve may spread and elicit a response in a muscle supplied by a different branch.
    - ○ Blink reflex studies may reveal synkinesis, which is not present in essential blepharospasm, dystonia, or seizures.
  - – Needle EMG shows irregular, brief, high-frequency bursts (150–400 Hz) of motor unit potentials, which correlate with clinically observed facial movements.
- Imaging studies:
  - – MRI is the neuroimaging study of choice, especially if an underlying space occupying compressive lesion is suspected.

6. *Interpret the MRI images* (► **Fig. 85.1**, ► **Fig. 85.2**, ► **Fig. 85.3**, and ► **Fig. 85.4**).
   - ► **Fig. 85.1** demonstrates a T2-weighted axial section MR image of the posterior fossa at the level of the internal auditory meatus. The image shows a flow void structure representing a blood vessel that is crossing and distorting the pathway of the facial nerve root exit.
   - ► **Fig. 85.2,** ► **Fig. 85.3**, and ► **Fig. 85.4** represent another patient with HFS. T1- and T2-weighted axial sections MR images of the posterior fossa at the level of the cerebellopontine angle (CPA) are seen in ► **Fig. 85.2** and ► **Fig. 85.3**. A tortuous basilar artery that is compressing the left CPA at the facial nerve exit root is visualized.
   - The patient had digital subtraction angiogram (DSA) of the vertebral artery (► **Fig. 85.4**) that confirmed the presence of dolichoectasia of the basilar artery.
   - One should keep in mind that not all HFS cases are idiopathic; and the aim of investigating the patient is to rule out an underlying pathology.
7. *What is the underlying etiology of HFS?*
   Etiological factors:
   - The actual cause of HFS is debatable.
   - The majority of cases of HFS are caused by an ectatic blood vessel that irritates the facial nerve by compressing or forming a loop around the nerve at the nerve exit zone. The usual offending arteries are the posterior inferior cerebellar artery complex, the anterior inferior cerebellar artery, or the vertebral artery.
   - A minority of cases of HFS are caused by venous compression.
   - Rarely, the condition may be secondary to facial nerve injury, facial nerve compression by a CPA tumor, regeneration of the facial nerve following facial palsy, or it may be a result of a brainstem lesion such as a stroke or a multiple sclerosis plaque.
8. *What are the surgical and nonsurgical therapeutic options?*
   - In mild and early cases, twitching can be controlled by the use of some antiseizure medications or minor tranquillizers, such as carbamazepine (Tegretol; Novartis, East Hanover, NJ), clonazepam, and diazepam. However, results are not always **satisfactory** and medications need to be taken on a long-term basis.
   - Botulinum toxin injection directly into the affected muscles can ablate the muscular spasm for several months, but its effect is temporary and the sensation of spasm often persists. Adverse effects include dry eyes, ptosis, eyelid and facial weakness, diplopia, and excessive tearing.
   - The response to the latter two treatment modalities varies and their effects often attenuate over time, necessitating a surgical treatment.
   - As for surgical management, the definitive procedure is a microvascular decompression (MVD). The offending blood vessel is mobilized from the nerve exit zone. This may be performed though microscopic, endoscopic, or a combined method. Use of endoscopic visualization can improve the overall outcome.
9. *Briefly describe your surgical procedure for HFS.*
   Steps of MVD:
   - Positioning and opening
     - – The surgery is done under general anesthesia.
     - – The patient is typically placed in either supine or the lateral decubitus position.
     - – Minicraniotomy inferior to the transverse sinus and medial to the sigmoid sinus is performed to expose the dura (2.0–2.5 cm in diameter; see ► **Fig. 85.5**).
     - – The dura is incised.
     - – Cerebrospinal fluid is drained slowly, allowing the structures of the posterior fossa to fall away without retraction.
   - Dissection and decompression
     - – Lateral or inferolateral cerebellar exposure of the CPA is undertaken.
     - – The acoustic-facial bundle is identified.
     - – The offending vessel is identified and, using microdissection and gentle manipulation, the adhesions and compressions from the vessel(s) on the facial nerve are lysed, and the nerve and vessel(s) are freed from one another (see ► **Fig. 85.6**).
     - – Small implants of shredded Teflon felt are placed to hold the vessel away from the cranial nerve root exit zone by changing the axis of the loop (see ► **Fig. 85.7**). Other techniques may include performing a dural sleeve to hold the artery away from the nerve, or glueing the artery to the posterior fossa dura.
     - – Veins are treated similarly or coagulated and divided.

## Answers (*continued*)

10. *What is the expected surgical outcome?*
    - Excellent results of complete or near-complete abolition of spasm can be seen in up to 93% of cases at 3 months after surgical decompression.
    - Long-term follow-up reveals more patients with total relief of their spasm.
    - Patients having reoperation should expect less favorable results: 61% complete or near-complete resolution of spasm.
    - If the patient still has spasm in the postoperative period, conservative follow-up of the patient will usually show progressive resolution of the residual spasm within the following month.
    - Neurosurgeons and the patients should be aware that delayed symptom disappearance after MVD for HFS is more common than it has been reported.

11. *What are the possible surgical complications?*
    General complication related to posterior fossa surgery specific to facial nerve MVD:
    - Partial or complete, temporary or permanent facial palsy may result from manipulation of the facial nerve.
    - Owing to the immediate proximity of the eighth cranial nerve, MVD of the facial nerve for HFS has a risk of producing ipsilateral hearing loss of various degrees. This may result from stretching the eighth cranial nerve between its exit from the brainstem and its entry into the internal auditory meatus as the surgeon places cerebellar retraction to expose the facial nerve root exit. This complication may be controlled if intraoperative brainstem auditory evoked potentials are monitored.

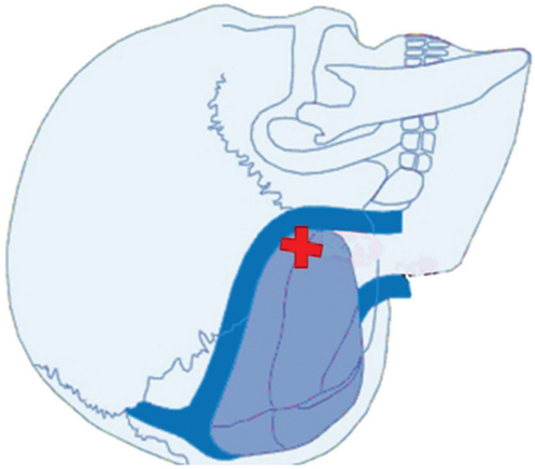

**Fig. 85.5** Illustration demonstrating head positioning in lateral decubitus. Location of craniotomy is marked by a *red cross*.

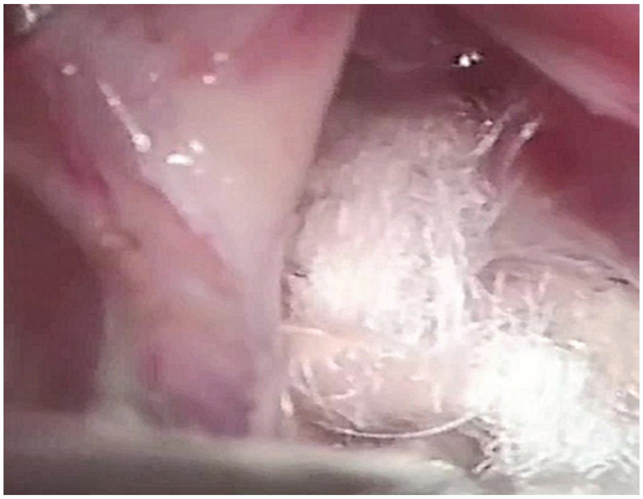

**Fig. 85.7** Intraoperative image demonstrating shredded Teflon felt placed to hold the vessel away from the cranial nerve root exit zone by changing the axis of the loop.

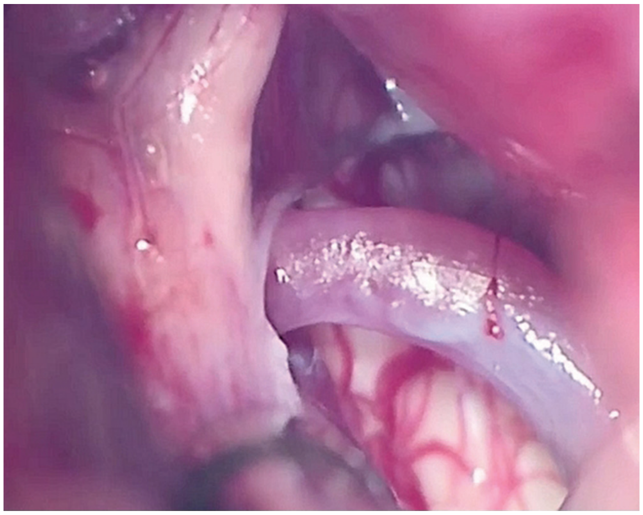

**Fig. 85.6** Intraoperative image demonstrating identification of an offending vessel along the facial nerve entry zone. Adhesions and compressions from the vessel on the facial nerve are lysed, and the nerve and vessel are freed from one another.

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# Case 86 Postherpetic Neuralgia

*Brian Gill and Christopher J. Winfree*

## Clinical Presentation

- A 75-year-old woman on chronic prednisone therapy for polymyositis presents 7 months previously with an outbreak of herpes zoster infection along her left abdomen and groin in a T12–L1 distribution.
- She experiences severe, intractable pain radiating "like a knife" within the distribution of the zoster rash.
- Although the rash gradually resolves, the pain persists, unchanged to the present.
- The pain grades an 8/10 in severity, is ameliorated partially with cold packs, and is exacerbated by light touch to the affected region.

## Questions

1. What is this patient's diagnosis and what measures could she have taken to prevent this condition?
2. What are the risk factors and pathophysiologic mechanisms underlying her condition?
3. What are her treatment options in the acute period (within 72 hours of rash onset)?
4. What are her treatment options in the chronic period (after the rash is resolved)?
5. What invasive treatment options are available if more conservative measures fail?

The patient undergoes a variety of pharmacologic treatments, including topical creams, oral anticonvulsants, antidepressants, and opiates, none of which yield acceptable pain relief. Multiple epidural steroid injections likewise failed to help. You are considering offering the patient either neurostimulation or a spinal infusion pump.

6. Describe the relative merits and drawbacks of each type of therapy.

## Answers

1. *What is this patient's diagnosis and what measures could she have taken to prevent this condition?*
    - Initially this patient had an acute herpes zoster outbreak in which a dermatomal outbreak is followed by a vesicular rash which gradually resolves over 30 days. Subacute herpetic neuralgia refers to pain which lasts up to 120 days after the initial rash.[1]
    - Pain lasting more than 120 days after the initial rash is known as postherpetic neuralgia (PHN) and occurs following 10 to 15% of acute zoster infections.[1]
    - Administration of the herpes zoster vaccine is a safe and efficacious method for preventing acute zoster and sequelae such as PHN. Vaccinated patients who develop zoster will still have a risk of developing PHN. However, zoster vaccine can significantly reduce morbidity from herpes zoster and PHN among older adults.[2]
2. *What are the risk factors and pathophysiologic mechanisms underlying her condition?*
    - Older age, female sex, a prodrome of dermatomal pain prior to rash appearance, and an immunocompromised state are risk factors for developing PHN.[3]
    - Sensitization and deafferentation are the major pathophysiologic mechanisms responsible for PHN. Reactivation of the virus leads to inflammation, tissue injury, and neuronal loss in both the dorsal root ganglion (DRG) and the periphery. Small unmyelinated C-fiber nociceptors become sensitized leading to reduced thermal sensory threshold and continual discharges.
    - Loss of C-fiber afferents in the DRG result in sprouting of A-beta fibers into superficial layers of the dorsal horn. This leads to connections between A-beta fibers and fibers which previously received input from C-fibers causing tactile allodynia.[4]
3. *What are her treatment options in the acute period (within 72 hours of rash onset)?*
    - Topical analgesics and oral anticonvulsants and antidepressants offer pain relief, but do not reduce the likelihood of developing PHN.
    - Oral antiviral agents administered within 72 hours of the onset of the acute outbreak may reduce the likelihood of developing PHN.[5]

## Answers (*continued*)

- The administration of oral corticosteroids in addition to oral antiviral agents during the acute period may reduce the duration of analgesic use in this period, but it does not prevent PHN.[5]

4. *What are her treatment options in the chronic period (after the rash is resolved)?*
   - Randomized, prospective clinical trials have shown that oral antidepressants, anticonvulsants, long-acting opioids, and nonsteroidal anti-inflammatory agents as well as topical anesthetic creams reduce pain in patients with PHN.[1]
   - Complementary and alternative medicine therapies such as physiotherapy and acupuncture have not been shown to be beneficial but may certainly be utilized with essentially no risk to the patient.
   - Transcutaneous electrical nerve stimulation (TENS) is a noninvasive method of neurostimulation often used in the management of chronic pain and it has been shown to have efficacy in PHN. Patches of electrodes placed over the painful area stimulate the skin beneath suppressing the nociceptive information transmitted by unmyelinated C-fibers via large diameter fiber stimulation.[6]
5. *What invasive treatment options are available if more conservative measures fail?*
   - Minimally invasive pain management techniques such as nerve blocks, sympathetic blocks, and epidural steroids have not been shown to be of benefit.[1]
   - Invasive surgical options such as neurectomy, cordotomy, dorsal root entry zone lesions, or other ablative therapies have been discredited and largely abandoned because of their risk of complications and lack of efficacy.[7]
   - Less invasive treatments such as intrathecal methyl prednisolone infusion may be helpful.[8]
   - Spinal cord stimulation, in which implanted electrodes in the spinal canal administer electrical impulses to the dorsal columns, may reduce pain and improve quality of life in PHN patients.[9]
6. *Describe the relative merits and drawbacks of each type of therapy*.
   - Spinal infusion pumps may offer dramatic pain relief for patients with chronic pain, including PHN. Implanting the devices is fairly straightforward for the practicing pain physician or neurosurgeon, and patients generally tolerate the systems fairly well. Patients generally do not perceive the effects of the infused drug(s), except for the relief of the pain.[10]
   - Nevertheless, the pumps need to be refilled every 1 to 3 months or so, and replaced after every 5 to 7 years. System malfunctions may occur prompting potentially lethal withdrawal syndromes in certain cases, and surgical revisions are sometimes required. Cerebrospinal fluid (CSF) leaks sometimes occur, following pump implantation or revision surgery.
   - In contrast, spinal cord stimulation systems generally require less maintenance than infusion pumps, there is no risk of a withdrawal syndrome if the system malfunctions, and CSF leaks are rare.
   - Stimulation produces noticeable paresthesias that are designed to overlap with the painful areas, eliciting pain relief. Some patients find these sensations annoying, especially if the paresthesias are in unwanted areas outside of the pain distribution.[11]
   - Spinal nerve root stimulation is a neurostimulation technique similar to spinal cord stimulation, except that the electrodes are placed more laterally in the spinal canal to preferentially stimulate the dorsal rootlets (▶**Fig. 86.1**). This provides similar levels of pain relief within the targeted painful areas as spinal cord stimulation, while limiting unwanted stimulation paresthesias to undesired areas.[11]
   - Deep brain stimulation can also be used to treat chronic pain syndromes including PHN. Stimulation of the contralateral ventral posterolateral (VPL) thalamic nucleus or the periventricular gray (PVG) at low frequencies can produce pain relief. The VPL is usually preferred as side effects of PVG stimulation may include nystagmus, oscillopsia, and impaired upgaze.[12]
   - Possible complications or drawbacks of neurostimulation implants include electrode displacement or malfunction, hematoma or pain at the site of pulse generator, requirement for battery replacement after 5 to 10 years.[11]

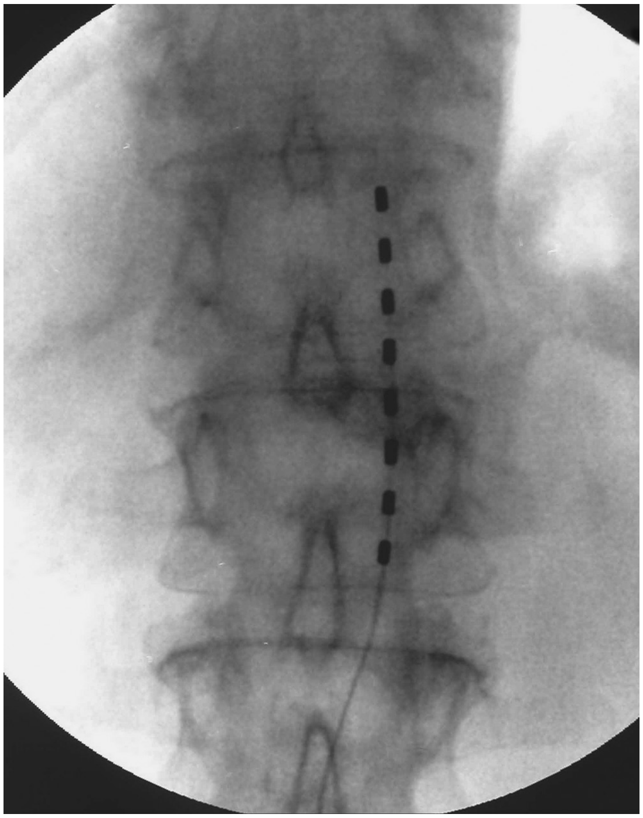

**Fig. 86.1** Intraoperative fluoroscopic image showing placement of a T12–L1 intraspinal nerve root electrode in the lateral aspect of the spinal canal. In this position, the electrical stimulation focused entirely upon the left-sided T12 and L1 dermatomes, the location of the patient's pain. There were no unwanted stimulation paresthesias in the lower extremity or elsewhere. The patient experienced significant (> 50%) and durable pain relief following placement of the spinal nerve root stimulator system and is quite satisfied with the result.

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# Case 87 Complex Regional Pain Syndrome in Children

*Flavio Giordano, Giuliana Rizzo, Anna Zicca, Remi Nader, and Lorenzo Genitori*

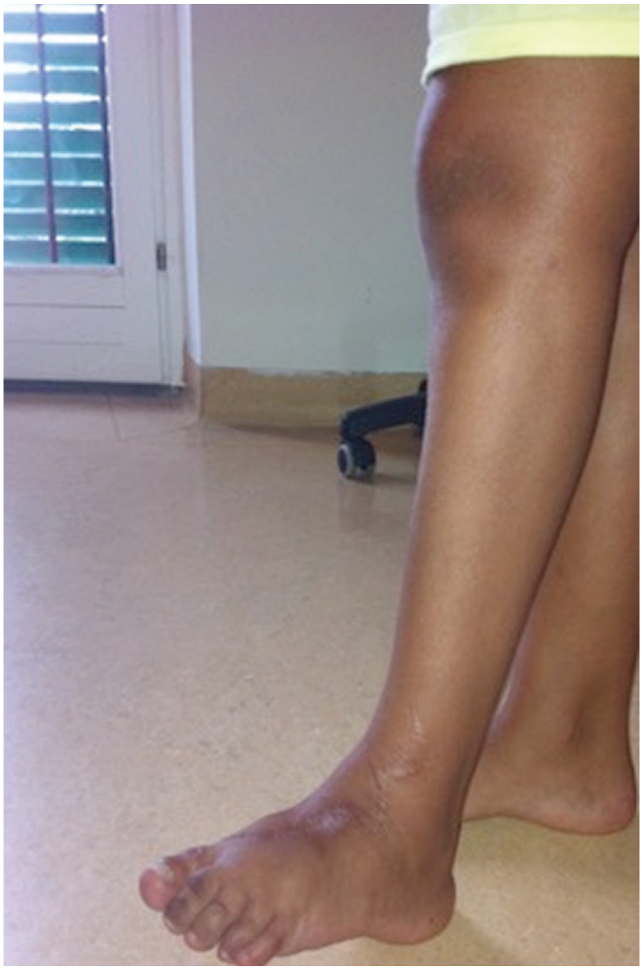

**Fig. 87.1** Severe motor impairment, patient unable to extend her left foot; trophic changes are clearly visible.

## Clinical Presentation

- An 11-year-old girl is referred to you by an orthopedic surgeon. She had a history of diffuse congenital angiomatosis in her left foot involving muscular structures. She has undergone multiple surgeries to reduce the angiomatosis since early childhood and has subsequently developed left foot pain refractory to medications, associated with hyperesthesia, edema, skin color and temperature changes (▶**Fig. 87.1**).
- The duration of her symptoms was 15 months. Due to the pain she was unable to extend her left foot and walk without crutches (▶**Fig. 87.1**). Boot immobilization was used for 40 days as an attempt to reduce pain.
- A MR-T2-weighted scan of the left foot showed diffuse angiomatosis localized into the plantar aspect of her foot, together with edema of the calcaneus and increased uptake after gadolinium enhancement.
- She was treated with nonsteroidal anti-inflammatory medications and opioids (tramadol) without pain relief.

## Questions

1. Based on the available information, what investigation would you perform?
2. What is the differential diagnosis for this child's pain?
3. What is the most likely diagnosis at this point?
4. What is the definition of complex regional pain syndrome (CRPS) according to the International Association for the Study of Pain (IASP)?
5. What symptoms and signs are associated with CRPS in children?
6. What are the possible neuropathological and neurophysiologic mechanisms of CRPS?
7. What treatment options can you offer in terms of multidisciplinary approach in children?
8. Describe the initial, noninvasive treatment of this condition.
9. Describe the invasive treatments of this condition if conservative measures fail.
10. What is the epidemiology of CRPS among the pediatric population?
11. A few years later, the patient still has recurrent pain in her foot. You now wish to place a spinal cord stimulator. Where will you position the electrodes?
12. What are the potential complications of spinal cord stimulation (SCS)?
13. What are the outcomes of SCS in patients with CRPS?

## Answers

1. *Based on the available information, what investigation would you perform?*
    - Electromyography with nerve conduction studies (EMG/NCS) may diagnose a peripheral nerve injury that was unidentified in the present case. This test should be performed at least 3 weeks after the initial injury to rule out denervational changes.
    - In children, EMG/NCS is generally difficult to perform, and most of the time there is no specific diagnostic laboratory test for the diagnosis of pain, requiring clinicians to rely predominantly upon sign and symptoms.[1]
2. *What is the differential diagnosis for this child's pain?*
    - Peripheral nerve injury
    - CPRS
    - Other left foot conditions that may be associated with pain upon presentation can include: infections (soft-tissue infections, osteomyelitis), inflammatory conditions, trauma to the foot or leg, vascular insufficiency, other types of neoplasms, iatrogenic causes secondary to the multiple procedures that she has sustained.
3. *What is the most likely diagnosis at this point?*
    - CPRS type I: This condition is divided into two categories based on the absence of a recognizable nerve injury (*CPRS type I, i.e., reflex sympathetic dystrophy*) or the presence of a proximal nerve injury (*CPRS type II, i.e., causalgia*).[2]
4. *What is the definition of complex regional pain syndrome (CRPS) according to the International Association for the Study of Pain (IASP)?*
    - CRPS is a term refined by IASP to describe disorders characterized by spontaneous or stimulus-induced pain that is disproportionate to the inciting event.
    - The disease is complex because it often includes a wide variety of autonomic and motor disturbance as abnormal blood flow, sweating abnormalities, and trophic change.[2,3]
5. *What symptoms and signs are associated with CRPS in children?*
    - Continued pain that is disproportionated to the inciting event.
    - A history of three of the following symptoms or signs[4,5]:
        - Sensory: hyperesthesia or allodynia
        - Vasomotor: temperature asymmetry, skin color changes, or skin color asymmetry
        - Sudomotor changes/edema: edema, sweating changes, or sweating asymmetry
        - Motor/trophic: decreased range of motion, motor dysfunction, or trophic changes
    - The presence of at least one of the following signs in two or more of the following categories:
        - Sensory: hyperesthesia or allodynia
        - Vasomotor: temperature asymmetry, skin color changes, or skin color asymmetry
        - Sudomotor changes/edema: edema, sweating changes, or sweating asymmetry
        - Motor/trophic: decreased range of motion, motor dysfunction, or trophic changes
        - Absence of another condition that could reasonably explain the findings
6. *What are the possible neuropathological and neurophysiologic mechanisms of CRPS?*
    - The exact mechanism of CRPS remains elusive.
    - Peripheral sensitization of A-delta and C afferent fibers to noxious stimuli appears to be the basis for CPRS-associated hyperalgesia.
    - In the spinal cord, there may be sensitization of a wide dynamic range neurons that occurs after intense peripheral stimulation of A-delta and C fibers.
    - The sympathetic nervous system is involved, especially when the pain or autonomic components are relieved using sympathetic blocks.

## Answers (*continued*)

- Central nervous system (CNS) alterations, probably in the cortex and thalamus, may also play a role in CPRS, especially in patients with extensive sensory deficits; however, it is not known if the CNS changes are primary or secondary to pain.[6,7]

7. *What treatment options can you offer in terms of multidisciplinary approach in children?*
   - Effective treatment of CPRS requires a multidisciplinary, pain management-oriented approach.[8–10]
   - Psychological evaluation and support is performed to rule out the existence of any psychological amplifiers of pain (i.e., behavioral problems, depression) and treat them if necessary, typically involving the family and school.[11,12]
   - Physical therapy enables recovery of function in the affected extremity and prevents disuse changes from occurring.
   - Pharmacological treatment is only a part of the therapy; it is helpful in children to temporarily reduce pain and facilitate motion, strength, and proprioception with sensory desensitization of the affected extremity.
8. *Describe the initial, noninvasive treatment of this condition.*
   - Initial options in children always involve a noninvasive approach. First-line pharmacological treatments include anticonvulsants and antidepressants. Opioids are a second-choice therapy, especially in case of "school refusal."[11,12]
9. *Describe the invasive treatments of this condition if conservative measures fail.*
   - If the noninvasive approach fails, anesthetic blocks of the peripheral sympathetic nerves and/or epidural analgesia may be considered under general anesthesia (▶**Fig. 87.2**).[13]
   - SCS is rarely applied in children.[14]
10. *What is the epidemiology of CRPS among the pediatric population?*
    - There is a paucity of large-scale studies about CPRS in children. In the recent years, there have been reports of an increase of CPRS in the pediatric population however.[15]
    - Applying the IASP criteria for CPRS, one study identified 74 cases of CPRS type I in a population of 106,470, resulting in a rate of 5.46 per 100,000 person years at risk, and a low period prevalence of 20.57 per 100,000.[15]
    - CPRS in the pediatric age occurs mostly in girls between 8 and 16 years of age, and is commonly localized in the lower limbs.[15]
11. *A few years later, the patient still has recurrent pain in her foot. You now wish to place a spinal cord stimulator. Where will you position the electrodes?*
    - The spinal cord stimulator electrodes can be either via percutaneous or laminectomy paddle leads (▶**Fig. 87.3**).
    - The leads should be placed in the dorsal epidural space around T12–L1, just off midline and eccentric to the left. Intraoperative fluoroscopy is used to determine the exact location.
    - The patient should experience stimulation paresthesias in the left foot, overlapping with the painful area.
    - This procedure is typically performed in the awake patient to confirm overlap and limit unwanted stimulation paresthesias to other parts of the body.
12. *What are the potential complications of spinal cord stimulation (SCS)?*
    - Displaced electrodes (21.5%)
    - Fractured electrode (5.9%)
    - Infection (3.4%)
    - Hardware malfunction (4.9%)
    - Subcutaneous hematoma (4.4%)
    - Discomfort over pulse generator (1.2%)
    - Cerebrospinal fluid leak (0.5%)[16,17]
13. *What are the outcomes of SCS in patients with CRPS?*
    - A meta-analysis of 25 case series with a median follow-up of 33 months found that 67% of CRPS patients achieved 50% or more pain relief.[18]
    - SCS is an appropriate and effective therapeutic option for patients with chronic benign pain refractory to medication or injection therapy.

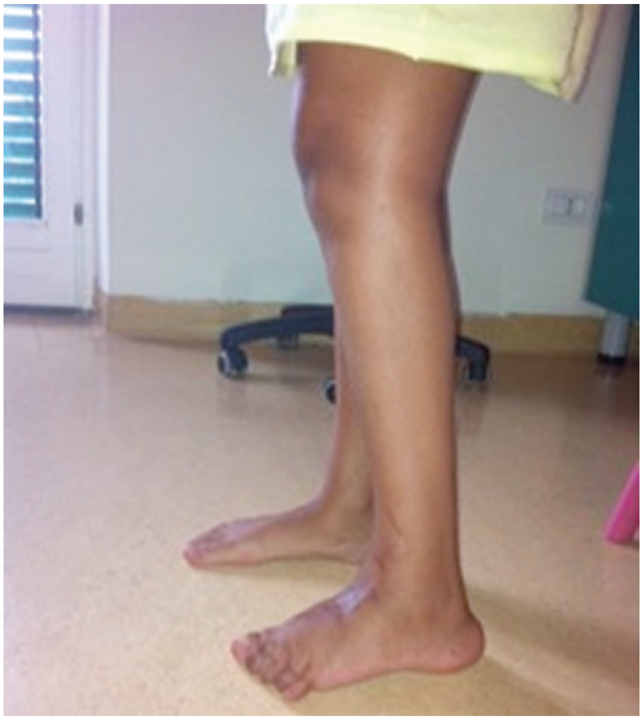

**Fig. 87.2** The child is able to move her foot after pain relief by peripheral sympathetic anesthetic block and manual rehabilitation.

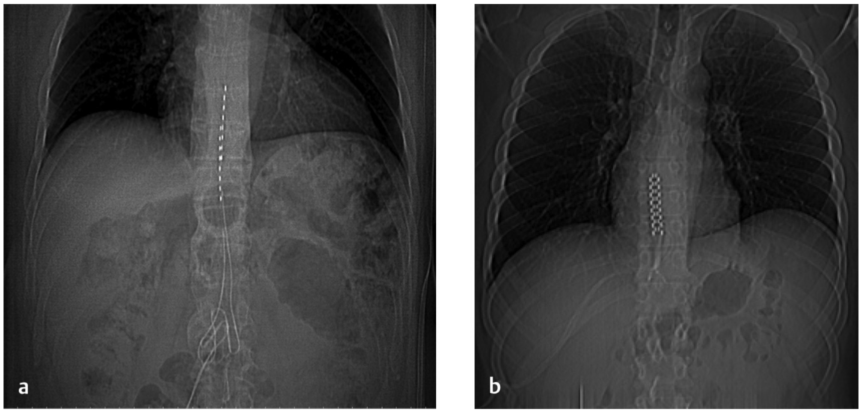

**Fig. 87.3** Examples of other patients with postoperative anteroposterior lumbar spine X-rays following placement of percutaneous leads (**a**) and laminectomy with paddle leads (**b**).

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# Case 88 Spasticity after Cord Injury

*Remi Nader*

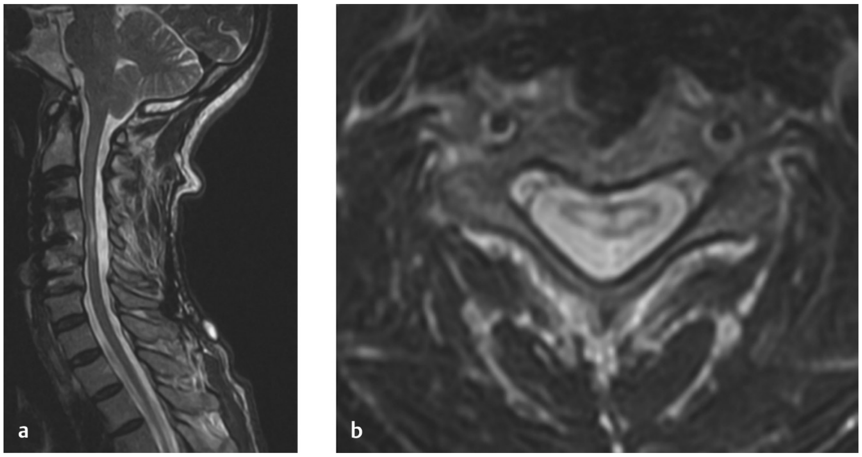

**Fig. 88.1** MRI of the cervical spine: **(a)** T2-weighted images with midsagittal section, and **(b)** axial section through C4–5 level.

## Clinical Presentation

- A 64-year-old man presents 6 years after having sustained a motor vehicle accident.
- The patient is a recovered C4 partial quadriplegic who underwent a C4 corpectomy and C5–6 anterior cervical diskectomy and fusion about 1 year ago.
- He now presents with some upper extremity pain and bilateral shoulder pain.
- He has some persistent spasticity in both upper and lower extremities, unsteady gait, and difficulties with bladder control.
- Hyperreflexia and muscle atrophy are seen diffusely on examination.
- He has been taking baclofen, tramadol, and diazepam, but the effect of the medications seems to have worn off despite a recent increase in dosage.
- MRI of the entire spine is done, and the only pertinent positive findings are shown in ▶**Fig. 88.1**.

## Questions

1. Interpret the MRI scan.
2. How do you manage the syrinx?
3. What are some other causes of spasticity?
4. Describe a grading system for spasticity.
5. What are some other medical options to treat spasticity in this patient?
6. Name five surgical options commonly employed to treat spasticity.
7. What are the main complications of pump placements?
8. What are the main selection criteria for placement of baclofen pumps?

## Answers

1. *Interpret the MRI scan.*
   - T2-weighted sagittal image (▶ **Fig. 88.1**a) shows a previous C3 to C6 anterior diskectomy and fusion.
   - Cord atrophy at the level of the upper cervical cord is noted.
   - A small syrinx is seen at the level of C4–5, but it is not causing any pressure on the cord or any compression of neural elements.
   - Cerebrospinal fluid (CSF) spaces are wide open around the spinal cord.
   - There is some straightening of the normal lordosis.
2. *How do you manage the syrinx?*
   - The syrinx is managed expectantly.
   - No treatment for the syrinx is needed as it does not cause any pressure on the spinal cord or neural elements.
3. *What are some other causes of spasticity?*
   - Multiple sclerosis
   - Cerebral palsy
   - Spinal dysraphism
   - Amyotrophic lateral sclerosis
   - Traumatic brain injury
   - Stroke
4. *Describe a grading system for spasticity.*
   - Ashworth grading system[1,2]:
     - 0: No increase in tone
     - 1: Slight increase in tone with small "catch" when moving affected limb
     - 2: More marked increase in tone with easy passive movements
     - 3: Significant increase in tone with hard passive movements
     - 4: Rigid affected part
5. *What are some other medical options to treat spasticity in this patient?*
   - Dantrolene[3]
     - Decreases calcium influx in sarcoplasmic reticulum
     - Decreases muscle contractions
   - Progabide[3]
     - GABA A and B activator
6. *Name five surgical options commonly employed to treat spasticity.*[1–3]
   - Baclofen and morphine pumps[4]
   - Electrical stimulation via epidural electrodes
   - Selective posterior rhizotomies[5]
   - Intramuscular phenol neurolysis
   - Myelotomies
   - Stereotactic thalamotomy
7. *What are the main complications of pump placements?*
   - Mechanical[4]:
     - Underinfusion
     - Catheter occlusion, kinking, dislodgment, or break
   - Wound problems[4]:
     - Erosion of pocket
     - Infection
     - Local pain
     - Seroma/hematoma
     - CSF collection
8. *What are the main selection criteria for placement of baclofen pumps?*
   - Selection criteria for baclofen pump are described below[4]:
     - Age 18 to 65 years, able to give informed consent
     - Severe chronic spasticity > 6 months
     - Spasticity refractory to oral medications
     - Ashworth scale at least 3
     - No CSF block
     - Positive response to intrathecal baclofen test dose
     - No implantable device
     - Nonpregnant patient
     - No allergy to baclofen
     - No history of stroke, renal insufficiency, severe liver or gastrointestinal disease

## Suggested Readings

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# Case 89 Neuronavigation and Intraoperative Imaging

*Lahbib A. Soualmi and Abdulrahman J. Sabbagh*

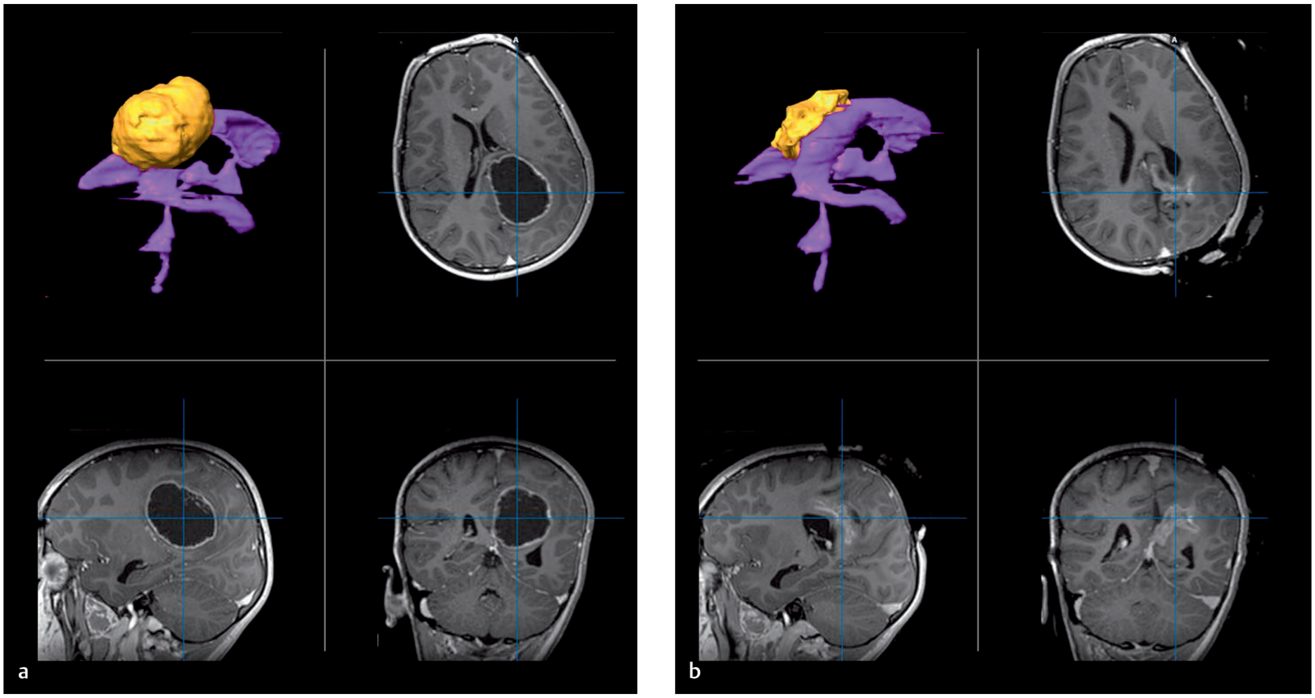

**Fig. 89.1** (**a**) MRI preoperative, three-dimensional (3D) reconstruction (*top left*) with ventricular system in *purple* and tumor in *yellow*. T1-weighted MRIs preoperative with contrast showing tumor in axial (*top right*), sagittal (*bottom left*), and coronal (*bottom right*) images. (**b**) MRI intraoperative, 3D reconstruction (*top left*) with ventricular system in purple and tumor in *yellow*. T1-weighted MRIs intraoperative with contrast showing tumor in axial (*top right*), sagittal (*bottom left*), and coronal (*bottom right*) images.

## ■ Clinical Presentation

- A 7-year-old right-handed girl presents with slowly progressive headaches and blurred vision.
- Examination is normal with the exception of early papilledema.
- MRI of the brain shows a large right parieto-occipital cystic lesion that is rim enhancing (▶**Fig. 89.1**).
- You decide to take the patient to surgery for diagnostic and therapeutic reasons.

## ■ Questions

1. How will you plan the surgery?
2. What are the characteristics of images needed for navigation in general?
3. What are the types of information that can be utilized for navigation?
4. How does neuronavigation work?
5. What are the types of registration available?
6. What are the existing types of localizers?
7. How do you classify intraoperative imaging?
8. Give examples where intraoperative imaging may be useful.
9. Compare the advantages and disadvantages of intraoperative CT (iCT) and intraoperative MRI (iMRI).

## Answers

1. *How will you plan the surgery?*
   - This patient has a cystic lesion. As the cyst leaks during surgery, the brain will shift and navigation will be very inaccurate. For this reason, the surgical team may decide to use intraoperative imaging and do the following (▶**Fig. 89.1**b):
     - Placing the head in a rigid MRI- or CT-compatible head holder in preparation for surgery.
     - Obtaining a preoperative MRI and calculating the cyst volume (▶**Fig. 89.1**a).
     - Performing craniotomy and frameless stereotactic aspiration of the cyst.
     - Performing iMRI after cyst aspiration to compensate for the brain shift caused by cyst aspiration and craniotomy (▶**Fig. 89.1**b).[1,2]
     - Taking an interhemispheric approach to the lesion and complete resection (▶**Fig. 89.2**).
2. *What are the characteristics of images needed for navigation in general?*
   - Three-dimensional (3D) global acquisition: Contiguous slices covering the whole region of interest such as the head or spine
     - Anatomic isotropic scan with high resolution: Fine cuts are required to be able to create an accurate volumetric data.
     - After computerized fusion, the functional data can then be superimposed on the anatomic data.
3. *What are the types of information that can be utilized for navigation?*
   - Anatomic information[3,4]:
     - MRI
       - T1-weighted ± contrast, T2-weighted, inverse recovery, fluid-attenuated inversion recovery sequences, etc.
       - Diffusion tensor imaging, tractography
       - MR angiography, MR venography
     - CT
       - CT ± contrast
       - CT angiography (CTA)
     - Ultrasound 3D acquisition
       - B-mode information
       - Doppler information
     - Conventional biplanar angiography
   - Functional information[5–8]:
     - Positron emission tomography (PET)
     - Single photon emission computerized tomography
     - Functional MRI (fMRI)
     - Magnetoencephalography (MEG)
     - Cortical electroencephalography
     - Depth electrode recordings
     - Transcranial magnetic stimulation
   - Biochemical information:
     - MR spectroscopy
     - Other chemical imaging modalities
4. *How does neuronavigation work?*
   - The following are the steps required for the workings of neuronavigation[9–12]:
     - Image acquisition (▶**Fig. 89.3**)
     - Image data analysis: Anatomical ± functional data analysis—segmentation and 3D reconstruction and volume rendering of objects of interest
     - Fusion and superimposition of different anatomical ± functional images (▶**Fig. 89.4**): The example given is PET data superimposed on MRI data.
     - Registration: Correlating and alignment of the 3D volumetric data with the actual patient (brain or spine)
     - Tracking: The neuronavigation platform should track the patient and surgical tools through the available localizer. This is done through different available tools (▶**Fig. 89.5**).

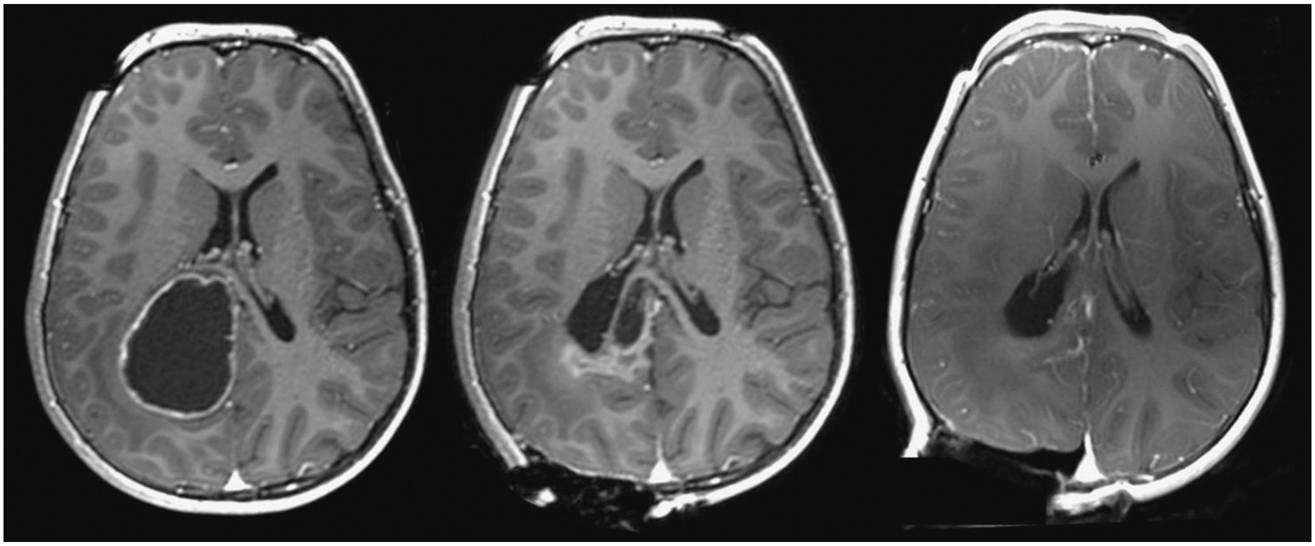

**Fig. 89.2** Intraoperative axial T1-weighted MRI with contrast showing progression of tumor resection.

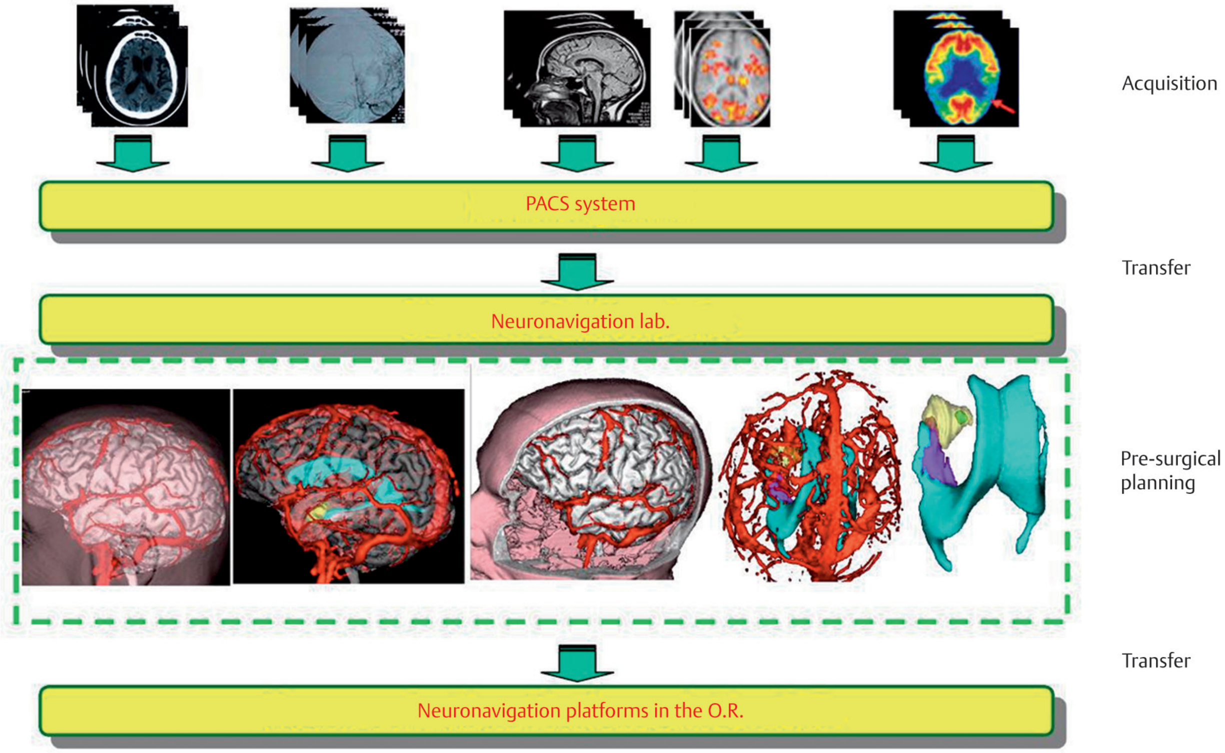
Acquisition
PACS system
Transfer
Neuronavigation lab.
Pre-surgical planning
Transfer
Neuronavigation platforms in the O.R.

**Fig. 89.3** Steps in neuronavigation (see text for further details). PACS, picture archiving and communication system; O.R., operating room.

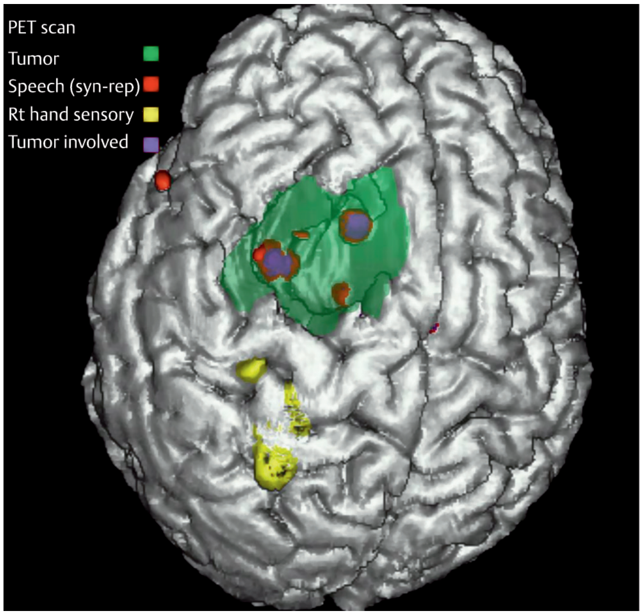
PET scan
Tumor 🟩
Speech (syn-rep) 🟥
Rt hand sensory 🟨
Tumor involved 🟪

**Fig. 89.4** Superimposed positron emission tomography scan with three-dimensional reconstructed MR images delineating the eloquent brain areas.

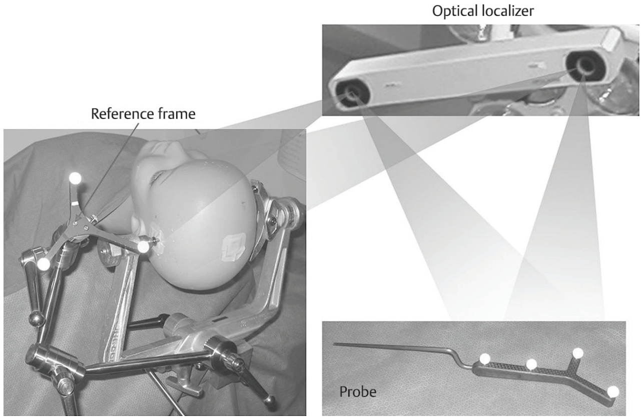
Optical localizer
Reference frame
Probe

**Fig. 89.5** Navigation process involving optical localization, frame referencing, and probe.

## Answers (*continued*)

5. *What are the types of registration available?*
    - Anatomic: Using anatomic landmarks (bridge of the nose, inner and outer canthi, tragus valleys, spinous processes, transverse process, etc.)[13]
    - Fiducials: Using radiopaque markers fixed before scanning the patient. These markers must be detected by the imaging modality used.[14]
    - Surface fitting: Pinpointing a mesh of points on the surface of the region of surgical interest using a probe or a laser scanning device to be correlated with the 3D object surface (i.e., face or spine of the patient).[13,15]
    - Automatic: Automatic recognition of geometric position of dedicated markers which are fixed to a head holder (in cases of iMRI) or scanner gantry (in cases of iCT) and scanned with the patient. They are used by the navigation platform to achieve the registration.
6. *What are the existing types of localizers?*
    - Optical localizers (using an infrared camera)[14]
        - Passive tracking: Using reflective spheres that are placed on the reference frame (patient), probes, and/or any surgical instrument (▶**Fig. 89.5**)
        - Active tracking: The reference frame (patient) and probes are equipped with light-emitting diodes.
    - Magnetic localizers: Magnetic signals that are emitted by the probe and picked up by a receiver indicating its location in space. This modality avoids the disadvantage of a clear line of sight required for optical localizers.[16]
    - Mechanical localizers: Articulated arms equipped with transducers giving the angular position
    - Ultrasound localizers: Navigation probes transmit ultrasonic signals that are picked up by a receiver indicating its location in space.
7. *How do you classify intraoperative imaging?*
    - Realtime[3,17]:
        - Intraoperative ultrasound (iUS)
        - Intraoperative angiography
        - Fluoroscopy
    - Preacquired[11]:
        - iMRI
        - iCT
8. *Give examples where intraoperative imaging may be useful.*
    - Here are some examples of the use of intraoperative imaging.
        - iMRI, iCT, or iUS for detection of residual tumor[18]
        - iMRI, iCT, or iUS to compensate for brain shift
        - Reregister using intraoperative landmarks if the patient moves during surgery
        - iMRA, iCTA, or intraoperative angiography after clipping aneurysms or resecting arteriovenous malformations from the brain or spinal cord[17,19]
        - iUS to localize ventricles, cysts, and lesions in the brain and spinal cord
        - iUS for recalibration of navigation
        - iCT or iMRI for immediate localization of depth electrodes, strips, and grids (in epilepsy surgery), or deep brain stimulators (in functional and behavioral neurosurgery) to readjust them if needed[20]
9. *Compare the advantages and disadvantages of intraoperative CT (iCT) and intraoperative MRI (iMRI)*
    - See ▶**Table 89.1**[21–24]

**Table 89.1** Advantages and disadvantages of intraoperative CT (iCT) and intraoperative MRI (iMRI)

| | iCT | iMRI |
|---|---|---|
| Need for nonferromagnetic tools | No | Yes (within the 5 Gauss line) |
| Radiation exposure to patient and medical staff | Yes | No |
| Soft tissue accuracy | + | +++ |
| Bony information | +++ | + |
| Angiography and venography | Yes | Yes |
| Functional imaging | No | Yes |
| Tractography | No | Yes |
| Need for different coils for different regions of the body | No | Yes |
| Imaging acquisition time | Seconds | Minutes |
| Cost | Less | More |

*Source:* Data from Archip et al 2008; Lipson et al 2001; Okudera et al 1994; Engle and Lunsford 1987.[21–24]

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# Case 90 Deep Brain Stimulation: Parkinson's Disease

*Weston T. Northam, Joshua Loewenstein, and Eldad J. Hadar*

## Clinical Presentation

- A 63-year-old man is referred to your clinic by a movement disorders specialist with medically intractable Parkinson's disease (PD).
- The patient was initially diagnosed with PD 5 years prior after he noticed a subtle pill-rolling tremor.
- He had a good initial response to medical therapy.
- Despite treatment with increasing doses of medication, the patient continues to experience long symptomatic intervals.
- The patient has begun developing involuntary writhing movements shortly after taking his medications, which make him socially withdrawn.
- An MRI of brain was obtained and was unremarkable.
- On physical examination off medication, the patient has significant upper extremity cogwheel rigidity, akinetic gait with shuffling steps, difficulty in initiating movement, and resting hand tremor.
- Minimal cognitive dysfunction or mood alterations have been noticed in this patient's case.

## Questions

1. What are the cardinal manifestations of PD? What secondary features can be found in PD?
2. Describe the neural circuitry that is affected in PD. How does the neurodegenerative process produce the constellation of symptoms seen in PD?
3. What pharmacologic therapies are available for patients with PD and what are the common side effects of these therapies?
4. When evaluating a patient with PD for deep brain stimulation (DBS), what are the primary considerations for selecting a good candidate? How can the severity of PD symptoms be objectively quantified?
5. What are the primary targets used for DBS in PD? Describe in detail the anatomy of these regions.
6. What tools do we have to confirm placement of the electrode in its defined target?
7. What side effects can be seen from high-voltage stimulation or improperly placed electrodes? Relate this to the anatomy of the DBS target.
8. What clinical result should the patient expect following activation of a properly placed DBS lead? When can they expect these changes to occur?
9. What are the steps to troubleshoot inefficacy of a DBS system postoperatively?
10. What complications are associated with DBS surgery? How prevalent are these complications?

## Answers

1. *What are the cardinal manifestations of PD? What secondary features can be found in PD?*
   - Bradykinesia, resting tremor (4–6 Hz), rigidity "cogwheeling," freezing, postural reflex loss[1]
   - Peak age of onset 60 years.[2]
   - Secondary features include: depression, anxiety, dementia, bradyphrenia (slowness of thought), akathisia, restless legs, autonomic dysfunction (hypotension, bladder/bowel dysfunction, sexual dysfunction), sleep dysfunction/fatigue.[1]
2. *Describe the neural circuitry that is affected in PD. How does the neurodegenerative process produce the constellation of symptoms seen in PD?*
   - Neuronal cell loss within substantia nigra pars compacta (60–70% loss at the ventrolateral tier at onset of symptoms)[2]
   - This results in loss of nigrostriatal dopamine, which normally is excitatory at D1 and inhibitory at D2 receptors (▶**Fig. 90.1**).
   - Due to the dopamine loss, there is resulting excitatory activity in the subthalamic nucleus (STN), globus pallidus interna (GPi), and inhibitory activity in the thalamus and motor cortex, which are thought to generate the motor symptoms such as bradykinesia, resting tremor, and rigidity.[1]
   - The STN and GPi undergo characteristic changes in neuronal firing patterns, rate, and oscillatory activity.[3]
3. *What pharmacologic therapies are available for patients with PD and what are the common side effects of these therapies?*
   - Most pharmacotherapeutic agents for PD have the end result of increasing dopaminergic transmission,

## Answers (*continued*)

either directly or indirectly. Side effects of these medications include motor overexcitation (dyskinesia) and hallucinations.

- The mainstay of treatment is levodopa (L-dopa) and decarboxylase inhibitor (carbidopa) that decreases L-dopa's peripheral breakdown.
- Catechol-O-methyl transferase (COMT) inhibitors (entacapone, tolcapone) are also used to achieve increase in dopaminergic transmission, as well as dopamine receptor agonists (cabergoline, bromocriptine, pergolide, ropinirole, and pramipexole).
- Monoamine oxidase B inhibitors (MAO-B), such as rasagiline and selegiline,[4] as well as anticholinergics and antiglutamatergics (amantadine) can provide ancillary support for parkinsonian symptoms.[1]

4. *When evaluating a patient with PD for deep brain stimulation (DBS), what are the primary considerations for selecting a good candidate? How can the severity of PD symptoms be objectively quantified?*
   - Thorough trials of multiple medication classes should be undertaken prior to consideration for surgery.
   - The following questions should be addressed during the selection process for DBS:
     - What is the patient's response to dopaminergic therapy?[3]
       - A good response to dopaminergic therapy and/or unacceptable medication side effects are generally predictive of good DBS outcome.
     - What is the severity of "on-off" fluctuations?
       - Severe on-off fluctuations are predictive of good DBS response.[5]
     - Is the patient's resting tremor medication-resistant?
       - Medication-resistant tremor is predictive of good DBS response.
     - Does the patient have underlying dementia or psychiatric disease?
       - Severe dementia and/or advanced psychiatric disease are contraindications.
     - Is there a history of atypical Parkinson's (progressive supranuclear palsy, multisystem atrophy, Lewy body dementia, Shy–Drager syndrome) or Parkinsonism secondary to medications/toxicity?
       - These are associated with poor response to DBS.[3]
   - Good family support at home is an important factor to assess.
   - Relative contraindications include: increased risk of intracerebral hemorrhage (anticoagulation or antiplatelet agents that cannot be stopped, uncontrolled hypertension), unstable cardiopulmonary disease, active infection.[6]
   - DBS should not be expected to help patients with predominantly nonmotor symptoms such as hypophonic speech, gait/postural instability, or secondary features of PD as mentioned above.
   - The Unified Parkinson's Disease Rating Scale (UPDRS-III) is a clinically validated tool used to assess the severity of motor symptoms, which can be used to measure the disease severity or gauge therapeutic efficacy.[7]
   - Mean improvement in UPRDS-III scores can range from 40 to 70%.[6]

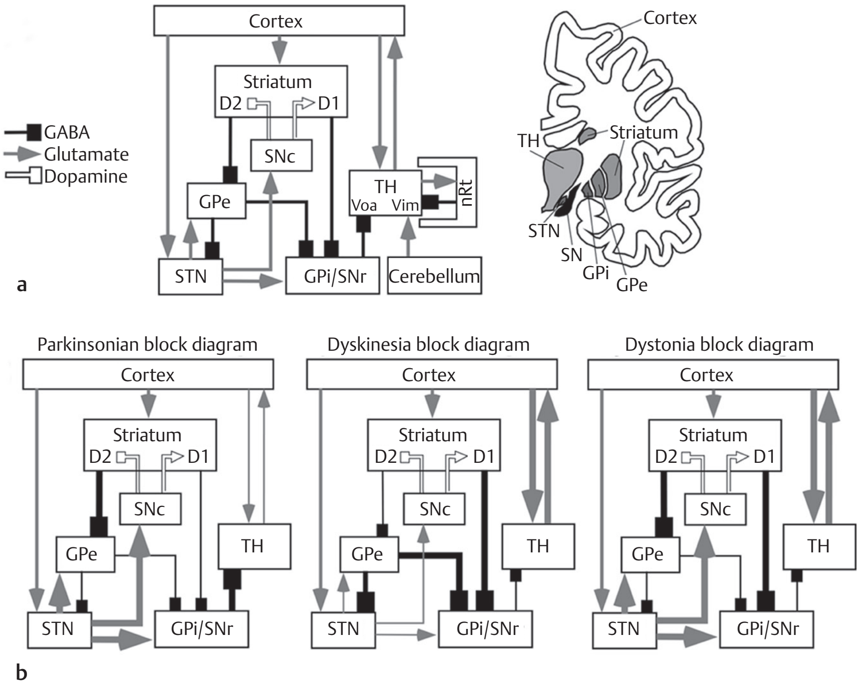

**Fig. 90.1** A schematic diagram of the excitatory and inhibitory interconnections between the basal ganglia in both the normal state (**a**), and in Parkinson's disease (**b**). c pars compacta; D dopamine; e externus; GP globus pallidus; i internus; r pars reticularis; SN substantia nigra; STN subthalamic nuclei; TH thalamus. (Reproduced with permission from Roy AE Bakay. Movement Disorder Surgery: The Essentials. Thieme; 2009, Fig. 2.1 a and b).

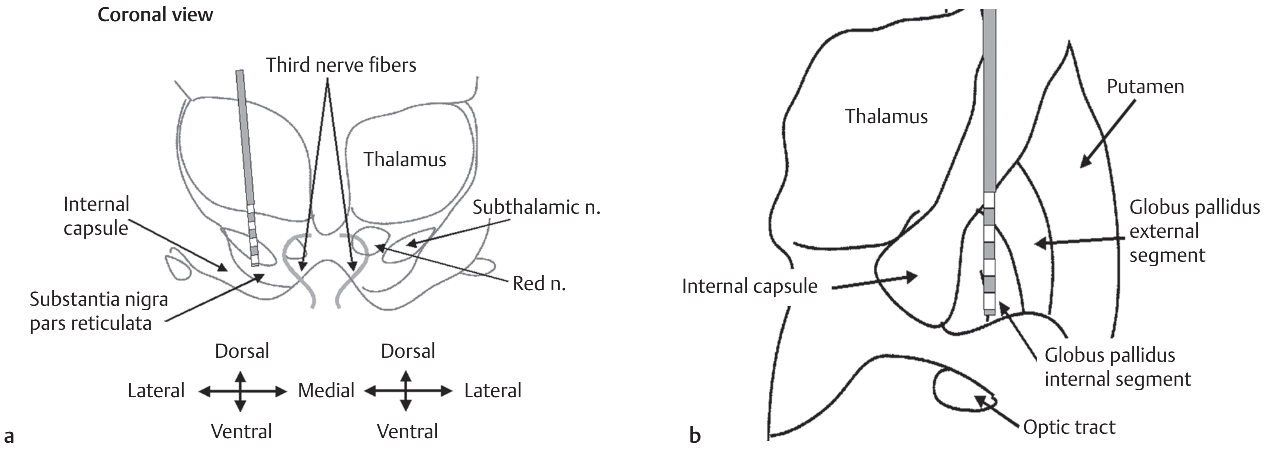

**Fig. 90.2** (**a**) A coronal view of a properly placed lead into the subthalamic nucleus, with the relevant surrounding anatomy. (**b**) A coronal view of a properly placed lead into the globus pallidus interna, with the relevant surrounding anatomy. (Reproduced with permission from Roy AE Bakay. Movement Disorder Surgery: The Essentials. Thieme; 2009, Fig 13.20 and 13.30).

## Answers (*continued*)

5. *What are the primary targets used for DBS in PD? Describe in detail the anatomy of these regions.*
   - Common stimulation targets in PD include the STN and GPi.
   - Ventral intermedius nucleus of the thalamus has been targeted but primarily has efficacy in treating the tremor component of PD.
   - The STN is a small lens-shaped structure of the basal ganglia located caudal to the thalamus. The boundaries of the STN include the zona incerta and ventral thalamus dorsally, the internal capsule anteriorly and laterally, the medial lemniscus posteriorly, the substantia nigra, third cranial nerve, and red nuclei ventrally, and the hypothalamus anteriorly (▶**Fig. 90.2**a).
   - The GP is another structure of the basal ganglia. The boundaries include the GP externa (GPe) and then putamen, anteriorly, dorsally, and laterally, the posterior limb of the internal capsule posteromedially, and the optic tracts ventrally (▶**Fig. 90.2**b).
6. *What tools do we have to confirm placement of the electrode in its defined target?*
   - Once a cannula is placed along the desired trajectory, a microelectrode recording can be performed.
   - Characteristic oscillatory patterns that are noted in both the STN and GPi can increase confidence that the electrode is properly placed.
   - Intraoperative test stimulation with awake and participating patient allows real-time feedback of the location of the electrode based on our understanding of the surrounding neuroanatomy. Gradual escalation of the stimulation amplitude can be trialed to create a larger stimulation field and elicit side effects.
   - Intraoperative imaging modalities, such as MRI and CT, can be used to assess the anatomic location of the stimulating electrode.
7. *What side effects can be seen from high-voltage stimulation or improperly placed electrodes? Relate this to the anatomy of the DBS target.*
   - An improperly placed lead can be defined radiographically or based on stimulation effects.
   - DBS leads that are suboptimally placed will likely produce stimulation side effects at relatively low levels of stimulation or would not produce expected benefits.
   - The nature of the side effects is consistent with the anatomic location of the improperly placed lead; refer to ▶**Table 90.1**, ▶**Table 90.2**, and ▶**Fig. 90.2**a, b.
8. *What clinical result should the patient expect following activation of a properly placed DBS lead? When can they expect these changes to occur?*
   - With stimulation of a properly placed lead in the STN or GPi, the patient can expect improvement in the cardinal manifestations of PD, i.e., rigidity, bradykinesia, and especially tremor.
   - Improvement of the postural or axial symptoms of PD can occur, but not as reliably as the other cardinal manifestations.
   - Typically, STN stimulation produces relief of tremor within seconds, rigidity within minutes, and bradykinesia within minutes to hours. The effect on axial symptoms often takes hours to days to occur.[9]
9. *What are the steps to troubleshoot inefficacy of a DBS system postoperatively?*
   - Postoperative alterations can be made to the neurostimulator settings to reshape the field that receives electrical stimulus. These changes help optimize effect and minimize adverse effects.
   - The settings that can be adjusted include amplitude, frequency, activation of different contacts

## Answers (*continued*)

along the electrode, and bipolar versus monopolar stimulation.

- Monopolar stimulation (use of the generator case as the anode) delivers electricity concentrically surrounding the chosen contact(s). Bipolar stimulation (anode and cathode both located on the stimulating electrode) delivers a more linear, ovoid electrical field along the axis of the electrode.
- Increasing the amplitude of stimulation expands the field that is stimulated.
- Pharmacotherapy, specifically dopaminergic therapy, should be titrated to effect in conjunction with changes made to the DBS system.

10. *What complications are associated with DBS surgery? How prevalent are these complications?*
    - Complications can be subdivided into three categories:
      – Surgery-related issues: Intracerebral hemorrhage (1–5%), seizure (3.1%), pulmonary embolism (0.4–4.9%), perioperative confusion (1–36%), suboptimal lead placement (3.8–12%)
      – Device-related issues: Infection (3–10%), skin erosion (2.5–6.45%), electrode or lead fracture (5.1–18%), lead migration (5.1%), neurostimulator malfunction/migration (18%)
      – Stimulation-related issues: Depends on the target and the direction in which the electrode is misplaced. See question 7.[8]

**Table 90.1** Orientation of the electrode lead relative to the STN can produce varying effects depending on whether the internal capsule, medial lemniscus/sensory thalamus, oculomotor fibers, or substantia nigra are stimulated

| Clinical Effect | Structure Stimulated | Anatomic Location Relative to STN |
|---|---|---|
| Tonic muscular contractions or dysarthria | Internal capsule | Lateral/anterior |
| Paresthesias | Medial lemniscus/sensory thalamus | Medial/posterior |
| Diplopia/visual disturbances | Oculomotor fibers | Medial/anterior |
| Affective changes | Substantia nigra | Inferior |

**Table 90.2** Orientation of the electrode lead relative to the GPi can create side effects if stimulation of the internal capsule or optic tract is encountered

| Clinical Effect | Structure Stimulated | Anatomic Location Relative to GPi |
|---|---|---|
| Tonic muscular contractions or dysarthria | Internal capsule | Medial/posterior |
| Visual disturbances | Optic tract | Ventral |

## Suggested Readings

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# Case 91 Deep Brain Stimulation: Essential Tremor

*Jonathon Lebovitz, Pratap Chand, and Richard Bucholz*

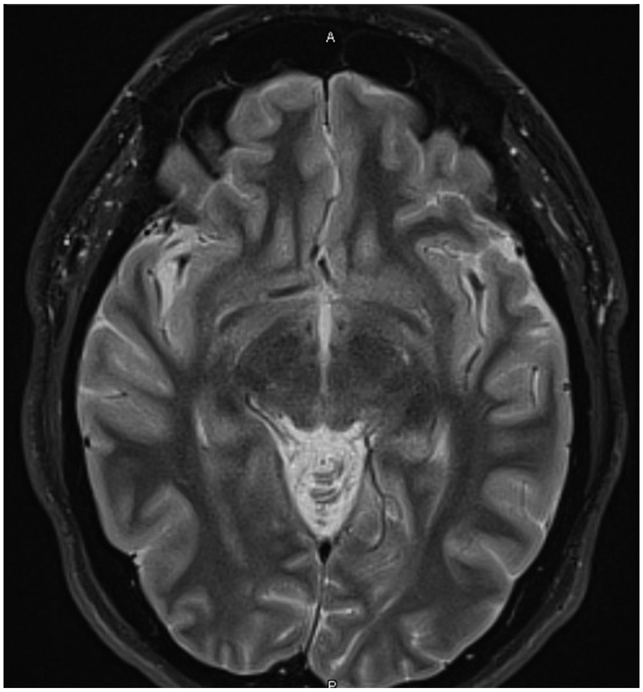

**Fig. 91.1** Preoperative image: MRI brain T2-weighted axial image through the ventral intermediate nucleus of the thalamus.

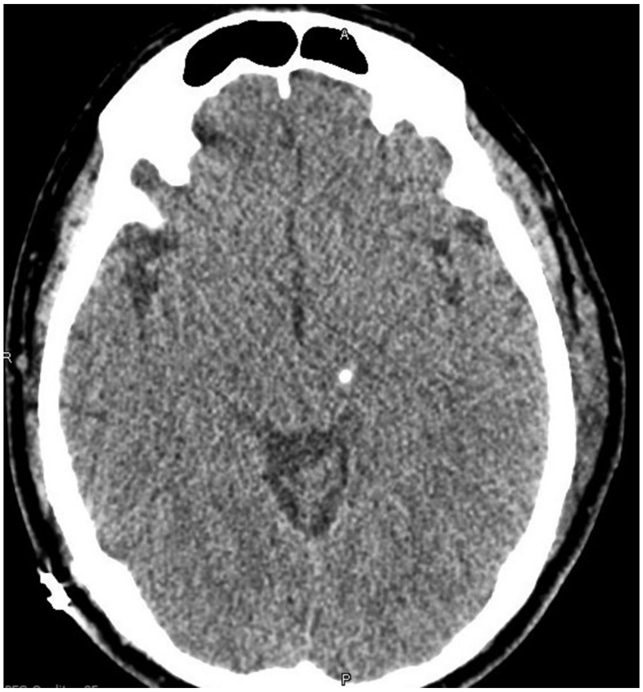

**Fig. 91.2** Postoperative image: CT scan of the brain, axial image showing the lead in the ventral intermediate nucleus of the thalamus. CT was performed on the first postoperative day.

## Clinical Presentation

- A 63-year-old man presents with a 20-year history of tremor in his upper extremities.
- The tremor has increased over the years and is severely disabling at this time where he cannot cook, write, use a computer, and spills his food.
- MRI of the brain did not show any significant abnormalities.
- He has tried multiple medications without complete relief of his tremor including propranolol, zonisamide, and primidone.
- His neurologic exam is significant for bilateral, asymmetric kinetic tremor without rigidity or bradykinesia.
- Work-up for Parkinson's disease is negative.
- The patient is otherwise healthy (▶**Fig. 91.1**).

## Questions

1. What is the most likely diagnosis?
2. What are the most common medications used to treat this disease?
3. What are the surgical treatment options?

   At this point, the patient wishes to undergo placement of a deep brain stimulator lead and an implantable pulse generator.
4. Where are the ideal lead placement locations to treat this condition?
5. To verify efficacy of lead placement, what intraoperative technologies can be employed to maximize outcome?
6. What findings would you see on intraoperative microelectrode recording (MER) or neurologic exam during ventral intermediate (Vim) nucleus of the thalamus lead placement if the lead is
   a. Lateral
   b. Anterior

## Questions (*continued*)

   c. Posterior
   d. Deep
7. What are possible surgical techniques for electrode implantation?
8. What is the likely outcome if deep brain stimulation (DBS) is successful? What is the average efficacy of lead placement?
9. What are the likely complications of this procedure?

   Three weeks after the procedure, the patient returns to clinic with redness over the chest incision that tracks up the neck. The patient also has a fever of 101 degree F and pain associated with the redness. The chest pocket appears full but the there is no drainage from the incision.

10. What is the likely diagnosis including microbiology and management for the current clinical situation?

## Answers

1. *What is the most likely diagnosis?*
   - Essential tremor (ET) is the most likely diagnosis. ET is the most common movement disorder with a prevalence up to 4% in the general population. This number increases with older patients.[1]
2. *What are the most common medications used to treat this disease?*

   Beta blockers, anticonvulsants, benzodiazepines, and antipsychotics have been tried with varying levels of success.
   - Beta blockers are used most commonly as first-line medical treatment. The most common and overall efficacious beta blocker used is propranolol.[1]
   - Primidone and topiramate are the most common anticonvulsants used in ET. When compared to propranolol, no significant difference was appreciated between primidone and propranol.[2]
   - Clozapine was found to have significant tremor reduction. Other medications found to have tremor reduction included olanzapine and quetiapine. In general, these are second-line treatments with clozapine being the most efficacious.
3. *What are the surgical treatment options?*

   The following injections and surgical options have been described:
   - Injections of botulinum toxin into the most prominently affected muscles have been used and found to offer mild improvement.
   - Thalamotomy of the ventral intermediate nucleus (Vim) has been performed via stereotactic radiosurgery that has resulted in improvement in contralateral tremor in drug-resistant ET. The durability of improvement following thalamotomy is less than that seen with DBS as lesions can "heal" over time decreasing their therapeutic impact.
   - Thalamotomy of the Vim has also been performed with transcranial focused ultrasound with similar benefits to that seen with radiosurgery.[3,4]
   - DBS with Vim lead placement was originally pioneered in 1987,[5] and is the most commonly utilized surgical procedure for medically refractory ET.
4. *Where are the ideal lead placement locations to treat this condition?*
   - Both the subthalamic nucleus (STN) and Vim have been described as helping to control tremor. One large series did describe that in patients over 70 years of age, the STN should be avoided secondary to side effects (see ▶**Fig. 91.2** and ▶**Fig. 91.3**).[6]
5. *To verify efficacy of lead placement, what intraoperative technologies can be employed to maximize outcome?*
   - During surgery, lead location can be verified anatomically with the use of intraoperative CT scanning, functionally with microelectrode recording performed with the patient awake, and trial stimulation of the electrode prior to closure of the wound.
6. *What findings would you see on intraoperative microelectrode recording (MER) or neurologic exam during ventral intermediate (Vim) nucleus of the thalamus lead placement if the lead is*
   *a. Lateral*
   *b. Anterior*
   *c. Posterior*
   *d. Deep*

   The following findings are observed based on improper lead positioning:
   a. If the lead is too lateral: tonic muscle contraction (internal capsule)
   b. If the lead is too anterior: muscle contraction (IC)
   c. If the lead is too posterior: paresthesia effecting ventralis caudalis (Vc)
   d. If the lead is too deep: dysarthria[7]
7. *What are possible surgical techniques for electrode implantation?*
   - A stereotactic frame has been conventionally employed to place electrodes, with registration performed with an N bar fiducial array. Alternatively, a frameless system (NextFrame; Medtronic, Minneapolis, MN) can be employed which enhances patient comfort during surgery as functional verification techniques require the patient to be awake during these sometimes quite prolonged proce-

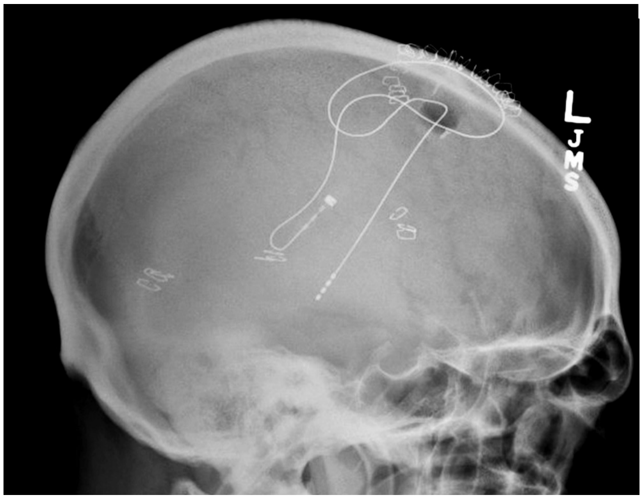

**Fig. 91.3** Lateral skull radiograph showing the lead in adequate position.

## Answers (*continued*)

dures.[8] Another frameless technique employs the use of a fabricated frame upon which to mount the microelectrode driving system (STarFix; FHC Inc.).[9] Finally, an MRI-compatible system has been created that allows electrodes to be anatomically tracked using an intraoperative MRI scanner.[10] Performing lead placement in an MRI precludes the ability to functionally locate the electrode using MER, however.

8. *What is the likely outcome if deep brain stimulation (DBS) is successful? What is the average efficacy of lead placement?*
   - Tremor improvement is the main successful outcome seen in ViM DBS surgery. The long-term published literature shows approximately 50% improvement in tremor. Activities of daily living (ADL) quality of life scores also ameliorated and have been found to be improved in up to 51% of patients.[11]
9. *What are the likely complications of this procedure?*
   - In addition to standard complications related to a cranial procedure, the following are also possible pitfalls: aborting the procedure, infection, bleeding, lead migration, malfunctioning pulse generator or lead, neurological deficits related to lead malposition or adjacent brain parenchymal damage such as weakness, loss of sensation, dysarthria, dysphasia, and seizures.[12,13]
10. *What is the likely diagnosis including microbiology and management for the current clinical situation?*
    - The patient is most likely presenting with a wound infection around the pulse generator. The patient should be taken to the operating room for removal of the generator and irrigation of the wound. The lead extension should also be removed from the contaminated area/explanted. After a course of both antibiotics, the system can be replanted into a new location. The intracranial lead is left in place.[14]

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# Case 92 Deep Brain Stimulation: Dystonia

*Faisal Al-Otaibi, Turki Elarjani, and Amal Mokeem*

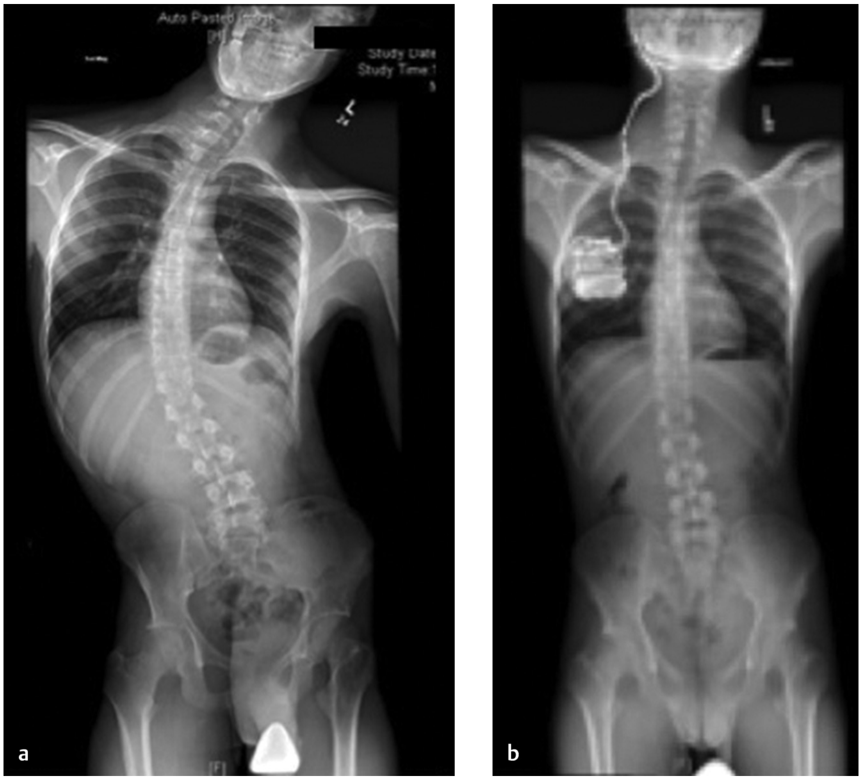

**Fig. 92.1** (**a**) X-ray showing full spine imaging in PA projection.
(**b**) X-ray showing full spine imaging in PA projection following deep brain stimulation placement.

## Clinical Presentation

- A 16-year-old boy presents with 1-year history of difficulty in writing with his right hand associated with involuntary abnormal movements in the right upper extremity.
- These abnormal movements continued to progress and later have been seen to involve other parts of the body. These are characterized by twisting movements of the upper extremities, difficulty in walking, and body tilt to the right side. He has been unable to hold objects, unable to run or participate in any sporting activities, and has had difficulties in standing.
- He reported that he can suppress the abnormal twisting movements in his body by holding his left arm with his right hand. His family history is positive for his cousin having a similar condition.
- He was evaluated in neurology clinic and found to have mutation of one allele of the *DYT1* gene (dystonia 1). A diagnosis of dystonia was made and the Burke–Fahn–Marsden Dystonia Rating Scale (BFMDRS) movement subscore was 82. His X-ray is shown ▶**Fig. 92.1**.

## Questions

1. What is the type of dystonia in this patient?
2. The patient described that he can suppress the dystonic movement by holding his left arm. What is the name of this phenomenon?
3. What is BFMDRS?
4. Describe the X-ray findings in ▶**Fig. 92.1**a.
5. Describe a classification of dystonia based on the distribution of the clinical features.
6. What are the types of dystonia based on etiology?
7. What are the commonly used initial treatments for dystonia?
8. What is the indication for deep brain stimulation (DBS) in dystonia?
9. What is the commonly used target for DBS for dystonia?

## Questions (*continued*)

10. What are the other alternative targets?
11. Define the X, Y, and Z Cartesian coordinates for globus pallidus internus (GPi) targeting in stereotactic surgery and describe the steps of the surgical technique.
12. Describe the potential side effects of stimulation based on the location of the DBS lead contact.
13. What is the outcome of DBS in such cases and in other types of dystonia?
14. What are the potential complications of DBS?

## Answers

1. *What is the type of dystonia in this patient?*
   - Primary dystonia
2. *The patient described that he can suppress the dystonic movement by holding his left arm. What is the name of this phenomenon?*
   - Sensory trick: A gesture or touching certain part of the body to suppress dystonic movement.
3. *What is BFMDRS?*
   - The BFMDRS is composed of two main subscales. Clinician-rated subscales:
     - A movement subscale, based on the patient's examination
     - A disability subscale, based on the patient's report of disability in activities of daily living
4. *Describe the X-ray findings in* ▶ **Fig. 92.1**a.
   - Full spine imaging has been obtained in the PA projection.
   - S-shape scoliosis is visualized with the upper curve convex to the right (apex at approximately T7–T9) and the lower curve is convex to the left. Coronal balance remains close to neutral (C7 spinous process to S1 spinous process).
5. *Describe a classification of dystonia based on the distribution of the clinical features.*[1]
   - Focal dystonia involves a single part of the body such as the hand.
   - Segmental dystonia affects one or more contiguous parts of the body.
   - Multifocal dystonia affects two or more noncontiguous parts of the body.
   - Hemidystonia involves one half of the body.
   - Generalized dystonia involves the entire body.
6. *What are the types of dystonia based on etiology?*
   - Primary (idiopathic) dystonia: can be inherited or sporadic
   - Secondary dystonia is associated with other neurological disorders such as neurometabolic disorders, vascular disorders, and trauma.
7. *What are the commonly used initial treatments for dystonia?*
   - Medication therapy such as baclofen, L-dopa, trihexyphenidyl, benztropine, tetrabenazine, tizanidine, clonazepam
   - Botulinum toxin injection
8. *What is the indication for deep brain stimulation (DBS) in dystonia?*
   - Patients with primary dystonia are more likely to have the best outcomes from DBS, and typically those who are younger, test positive for the *DYT1* dystonia gene mutation, and are treated relatively early on in the dystonia continuum.
   - Patients with severe cervical dystonia or dystonia acquired by drug exposure (tardive dystonia) may also be reasonable candidates for DBS.
   - Other patients with secondary dystonia should be evaluated on a case-by-case basis.
   - Typically, DBS can be considered if medications and other conservative treatments have failed, and if the symptoms negatively affect the quality of life to the extent that the surgical risks are justified.[2]
9. *What is the commonly used target for DBS for dystonia?*
   - Posteroventral GPi[3]
10. *What are the other alternative targets?*
    - Thalamus, subthalamic nucleus, and zona incerta
11. *Define the X, Y, and Z Cartesian coordinates for globus pallidus internus (GPi) targeting in stereotactic surgery and describe the steps of the surgical technique.*
    - GPi stereotactic coordinates are based on the mid-commissural point (MCP): X = 19–21 mm lateral to midline, Y = 2–3 mm anterior to MCP, and Z = 4–5 mm below the anterior commissure (AC)–posterior commissure (PC) plane. **Table 92.1** describes the stereotactic coordinates for different common targets for DBS in general.
    - The surgical techniques and steps are variable between centers. The authors describe the principles of the more commonly applied techniques.
    - Most surgeries are performed under local anesthesia, although in certain cases, such as pediatric patients and some dystonia cases, general anesthesia is used.[4]
    - The surgical steps start with the placement of a stereotactic frame to the head (see ▶ **Fig. 92.2**a).
    - An MRI of the brain is obtained to calculate the X, Y, and Z coordinates of the target with the aid of a planning station. Some centers use a CT of the brain that is then fused with preoperative MRI. Surgical planning software is helpful to correct misalignments of the frame, identify a cortical entry point that will avoid the sulci and ventricle, and aid in target localization.[5]

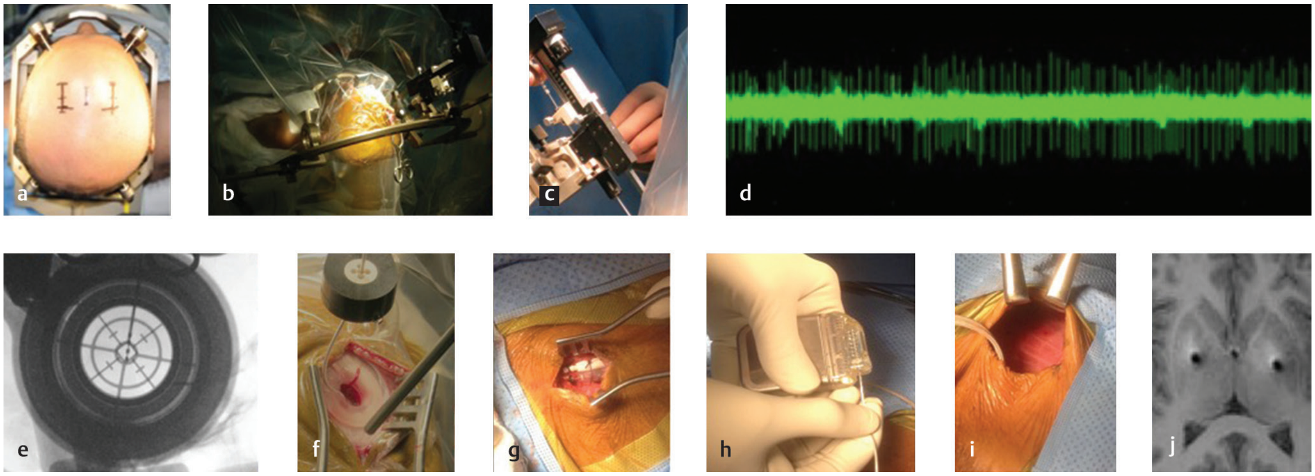

**Fig. 92.2** (**a**) Placement of Leksell frame preoperatively and marking of the skin incisions bilaterally. (**b**) Intraoperative picture showing placement of Leksell ark apparatus which is adjusted based on computer generated coordinates to target and entry point. (**c**) Depth electrode advancement to target using an anchor bolt attached to the arc apparatus. (**d**) Monitoring of electrophysiological responses at the level of the GPi. (**e**) Intraoperative fluoroscopy showing depth electrode cannula (track marker). Cannula creates a track for implantation of depth electrode. (**f**) Fixation of the deep brain stimulation lead after removal of inner stylet. (**g**) Plastic connector placed to protect the lead and maintain the anchoring. (**h**) Distal connection of the lead to the internal pulse generator (IPG). (**i**) Creation of a subclavicular pocket for implantation of the IPG. (**j**) Postoperative axial MRI showing proper placement of the leads within the GPi.

## Answers (*continued*)

- During the procedure, the Leksell frame base is attached to the operating table. The skin opening is typically located anterior to the coronal suture and lateral to the midline by at least 2 cm and is made after the skin has been infiltrated with local anesthesia.
- A burr hole is placed either at the planned cortical entry point or in a standard location such as Kocher's point (1 cm anterior to the coronal suture and 2–3 cm lateral to the midline).
- Neurophysiological verification is used to refine the final position of the DBS lead. This neurophysiological monitoring includes microelectrode recording (MER) and macrostimulation. Certain centers do not use neurophysiological verification methods.
- MER during GPi targeting is used to identify the globus pallidus externus (GPe) and the GPi. After passing the GPi, the optic tract is identified.
  - Stimulation at low intensity can evoke phosphenes or flashing lights in the contralateral visual field.
  - The GPi usually fires in high-frequency discharges, typically around 60 to 90 Hz. The GPi firing rate in dystonic patients may be lower than that in patients with Parkinson's disease (see ▶ **Fig. 92.2**d).
  - The final DBS distal contact is placed 1 mm above the optic tract and 3 mm anterior to the internal capsule.
- Macrostimulation is done to simulate the clinical effect and make sure that the lead position is optimal (see ▶ **Table 92.2**).
- In general, the DBS electrode is implanted with the guidance of a fluoroscope to verify its location and to rule out any deviation.
- The internal pulse generator (IPG) and its extension leads can be implanted during either the same procedure or a subsequent procedure (see ▶ **Fig. 92.2**h, i). This is carried out under general anesthesia with the patient in the supine position with his/her head turned contralaterally. The IPG is usually placed in the infraclavicular area, about 2 cm below the clavicle and 3 cm from the lateral manubrial border. Depending on the thickness of the skin and subcutaneous tissue, the IPG is placed either subcutaneously or under the pectoralis muscle fascia.

12. *Describe the potential side effects of stimulation based on the location of the DBS lead contact.*
    - Side effects of stimulation are summarized in ▶ **Table 92.2.**[6,7]
13. *What is the outcome of DBS in such cases and in other types of dystonia?*
    - DBS usually benefits patients with primary dystonia. Patients with a *DYT1* gene mutation respond better to DBS.
    - In contrast, secondary dystonia responds less to DBS than primary dystonia.
    - The improvement in primary dystonia after bilateral GPi DBS was reported to be around 60 to 80%.[8]
14. *What are the potential complications of DBS?*
    - Infection
    - Intracranial bleed
    - Seizure
    - Stimulation side effects (▶ **Table 92.2**)
    - Hardware failure

**Table 92.1** Commonly used stereotactic coordinates of selected targets for DBS[6]

| Target | X | Y | Z |
|---|---|---|---|
| **STN** | 11–13 mm lateral to midline | 3–4 mm (posterior) | 4–5 mm below AC–PC plane |
| **GPi** | 19–21 mm lateral to midline | 2–3 mm (anterior) | 4–5 mm below AC–PC plane |
| **Vim** | 11–12 mm lateral to the third ventricular wall | Midpoint between MCP and PC | At AC–PC plane |
| **Vc** | 2–3 mm anterior to PC | 12–13 mm from midline for facial pain<br>14–15 mm for upper limb pain<br>16–17 mm for lower limb pain | At AC-PC plane |
| **PVG** | 2–3 mm anterior to PC | 2 mm lateral to the medial third ventricle wall | At the level of AC–PC plane |
| **AN** | 6 mm lateral to MCP | 8 mm anterior to PC | 12 MM above AC–PC plane |

*Abbreviations:* AN, thalamic anterior; GPi, globus pallidus internus; PVG, periventricular gray matter; STN, subthalamic nucleus; Vc, thalamic ventrocaudalis; Vim, ventral intermediate nucleus of thalamus.

**Table 92.2** Correlation between stimulation induced clinical effects and the likely electrode sites in selected DBS targets

| Target | Electrode Sites | Stimulation Induced Effects |
|---|---|---|
| **STN** | Posteriorly or medially located electrode | Persistent paresthesia at low amplitude (stimulation spread to medial lemniscus) |
| | Lateral or anteriorly located electrode | Tonic contraction and dysarthria at low stimulation amplitude (internal capsule stimulation effect) |
| | Medial and anteriorly located electrode | Diplopia (oculomotor effect) |
| | Potentially too deep or medial electrode location | Adverse effect on mood |
| | Electrode location is above or too anterior to the STN | Absence of adverse effect and benefit |
| | STN location | Dyskinesia |
| **Vim** | Posteriorly located electrode | Persistent paresthesia at low amplitude |
| | Laterally located electrode | Dysarthria or tonic contraction (internal capsule stimulation effect) |
| | Electrode location is above or too anterior to the Vim | Absence of adverse effect and benefit |
| **GPi** | Deep location | Visual response (i.e., phosphenes) (stimulation spread to optic tract) |
| | Posterior or medially located electrode | Dysarthria or tonic contraction (internal capsule stimulation effect) |
| | Electrode location is above or too anterior or lateral to the GPi | Absence of adverse effect and benefit |

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# Case 93 Temporal Lobe Epilepsy

*Soha Abdu M. Alomar, Ashwag Al-Qurashi, Afnan Uthman Alkhotani, and Abdulrahman J. Sabbagh*

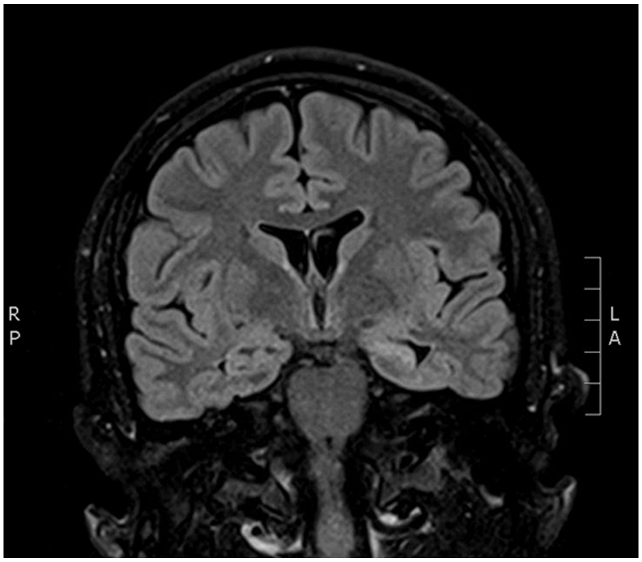

**Fig. 93.1** Fluid attenuated inversion recovery (FLAIR) coronal magnetic resonance image of the brain.

## Clinical Presentation

- A 28-year-old left-handed woman with history of febrile seizure as an infant presented with epilepsy diagnosis since childhood.
- She suffers from two types of seizures:
  - The first type is described as aura of a rising abdominal sensation and fear followed by loss of awareness associated with lip smacking and fine hand-motor automatisms. This is followed by postictal tiredness (2–3 per day).
  - The second type consists of nocturnal convulsions (2–3 per week).
- She was tried on several antiepileptics, and currently her epilepsy is refractory to triple medications.

## Questions

1. How do you classify her seizures?
2. Where would you localize her first type of seizures?
3. What would be your presurgical management steps?

   She had video electroencephalography (EEG) monitoring that showed ictal and interictal evidence of a left temporal focus. An MRI scan of the brain is performed (▶**Fig. 93.1**). Neuropsychological evaluation reveals normal intelligence, verbal and visuospatial memory. She was shown to be left hemisphere dominant, and Wada test lateralized her verbal functions, memory, and speech to the left side with no functional reserve in the right hemisphere.
4. Describe the finding shown on the MRI.
5. What baseline neuropsychological assessment carries more favorable outcome for left temporal lobectomy? What surgical options are available for this patient?
6. What are the chances of seizure control after anterior thalamic nucleus deep brain stimulator (DBS) placement, responsive neurostimulation system (RNS NeuroPace, Mountain View, CA) placement, or vagal nerve stimulator (VNS) placement?
7. If this patient was right sided dominant for language and memory, what are the surgical |options?
8. What is the expected seizure outcome after temporal lobe surgery compared with best medical management?
9. What is the chance of her becoming completely seizure free (Engel class Ia) after successful surgery? What are the chances of being on monotherapy after surgery? What are the chances of antiepileptic drug freedom after surgery?

## Questions (*continued*)

10. What are the possible complications associated with temporal lobe epilepsy surgery?
11. After performing left temporal lobectomy, the patient awoke with right body hemiplegia, what are the possible causes of this finding?
12. Describe the surgical principles in temporal lobe epilepsy surgery.
13. What is the central point? How does it help you during temporal lobe resection?
14. What is Meyer's loop and how can it be avoided during temporal lobe surgery?

## Answers

1. *How do you classify her seizures?*
   - The first type is a complex partial seizure.
   - The second type is probable secondary generalized tonic–clonic seizure.
2. *Where would you localize her first type of seizures?*
   - The semiology localizes these seizures to the temporal lobe or possibly the insula.
3. *What would be your presurgical management steps?*
   - Management steps include the following:
     - Obtaining a full history and clinical examination
     - Verifying antiepileptic medication serum levels
     - Obtain EEG followed by video EEG telemetry if the EEG was not conclusive
     - High-resolution MRI of the brain (1.5T or 3T)
     - Neuropsychological evaluation
     - Functional MRI (fMRI) for language lateralization
     - Wada testing (Dr. Juhn Wada's intracarotid sodium amobarbital procedure to lateralize speech and memory functions[1])
     - Other tests that could be performed for further lateralization and localization if above studies are still nonconclusive include: magnetoencephalogram (MEG), interictal positron emission tomography (PET), and ictal single-photon emission CT (SPECT).
4. *Describe the finding shown on the MRI.*
   - ▶**Fig. 93.1** shows a hyperintense signal and atrophy in the left hippocampus greater than on the right side.
   - These are characteristic features of mesial temporal sclerosis (MTS).
5. *What baseline neuropsychological assessment carries more favorable outcome for left temporal lobectomy?*
   - Patients with low average or low baseline verbal memory carry lower risk for postoperative decline in verbal memory.[2–4]
6. *What surgical options are available for this patient?*
   Since this is the dominant temporal lobe confirmed by the fMRI and Wada test with good baseline neuropsychological evaluation, surgical resection carries a high risk of postoperative neuropsychological decline; stimulation could be a good treatment modality. Options include:
   - RNS; NeuroPace (Mountain View, CA) implanted in the left hippocampus
   - Bilateral anterior thalamic nucleus stimulation (with DBS)
   - Vagal nerve stimulation
7. *What are the chances of seizure control after anterior thalamic nucleus deep brain stimulator (DBS) placement, responsive neurostimulation system (RNS NeuroPace, Mountain View, CA) placement, or vagal nerve stimulator (VNS) placement?*
   - The SANTE trial showed that after anterior thalamic nucleus stimulation, at 3 months, 54 patients who received stimulation had 40.4% seizure reduction while only 14.5% of the sham group had seizure reduction. By 2 years, there was a 56% median percent reduction in seizure frequency; 54% of patients had a seizure reduction of at least 50%, and 12.7% patients were seizure-free for at least 6 months.[5,6]
   - For RNS, NeuroPace placement, at 3 months, 97 patients who received the stimulation had 37.9% reduction while only 17.3% of the sham group had reduction. The responder rate at 1 year post implant was 43% and for those subjects who had reached 2 years post implant, it was 46%.[7]
   - With VNS, 35% of patients had reduction of at least 50% of seizure frequency and intensity; 20% showed 75% reduction. All stimulation studies show evidence of increasing response as the duration of stimulation increases.[8]
8. *If this patient was right sided dominant for language and memory, what are the surgical options?*
   As all seizures are coming from the left temporal lobe along with evidence of left-sided MTS, the surgical options are as follows:
   - Corticoamygdalohippocampectomy (temporal lobectomy)
   - Selective amygdalohippocampectomy
     - Transsylvian (Yaşargil technique)[9]
     - Transcortical (Olivier technique)[10]
     - Subtemporal
9. *What is the expected seizure outcome after temporal lobe surgery compared with best medical management?*
   - According to Wiebe et al,[11] (randomized clinical trial comparing temporal lobectomy with best medical management) the number of patients needed to be treated for one patient to become free of disabling seizures is two.
   - Fifty-eight percent of surgical cases compared with 8% of best medical management cases will be free of disabling seizures.[11]
   - Long-term favorable seizure outcome (Engel class I and II) ranges between 50 and 90%.[12,13]

AU1

## Answers (*continued*)

10. *What is the chance of her becoming completely seizure free (Engel class Ia) after successful surgery? What are the chances of being on monotherapy after surgery? What are the chances of antiepileptic drug freedom after surgery?*
    - According to the McGill group, 40 to 58% of patients undergoing temporal lobe epilepsy surgery end up in the Engel Ia class—seizure free.[13]
    - The chance of becoming antiepileptic drug free according to Téllez-Zenteno et al is 20%.[14]
    - Based on the same study, 41% will be on monotherapy.[14]
11. *What are the possible complications associated with temporal lobe epilepsy surgery?*
    - Complications vary among centers and range between 5 and 10%.[11] These include:
      - Infections
      - Hematoma
      - Hemiparesis
      - Memory and language deficits
      - Contralateral upper quadrantanopia— pie-in-the-sky deficit (Some consider it to be an expected finding rather than a complication.)
12. *After performing left temporal lobectomy, the patient awoke with right body hemiplegia, what are the possible causes of this finding?*
    - Vascular cause:
      - Arterial infarct: injury to anterior choroidal artery or middle cerebral artery (MCA) branches
      - Venous infarct: injury to vein of Labbe
    - Structural cause
      - Injury to cerebral peduncle/midbrain/internal capsule
      - Hematoma: intracerebral hemorrhage (ICH), epidural hemorrhage (EDH)
    - Functional cause
      - Postictal
      - *Describe the surgical principles in temporal lobe epilepsy surgery*.

        The steps in nonselective temporal lobectomy are as follows[9,10,15–17] (There are several surgical variations for temporal lobes epilepsy. Please refer to ▶**Table 93.1** for further details and for pros and cons for each one):
    - Positioning:
      - The aim is to have the frontotemporal area almost horizontal.
      - Patient is supine, with head in pins and roll under the shoulder.
      - The head is kept higher than the heart and turned 60 degrees, slightly extended, and exposing the temple.
      - Make sure the neck veins are not compressed or overstretched.
    - Craniotomy:
      - Adjuncts: neuronavigation is used to tailor the skin incision and bony opening, guide the trajectory to the temporal horn entry point, and verify the extend of posterior hippocampal resection.
      - Intraoperative imaging if available, such as iMRI or iCT, is helpful to confirm completion of resection or for reregistration after brain shifts.
      - Reversed question mark skin incision from the zygoma just anterior to the ear going back around the pinna and up and parallel to the temporalis line to the hairline is made.
      - Temporalis fascia and muscle opening are completed.
      - Exposure of the pterion and the zygomatic root is performed.
      - The craniotomy should reach the anteroinferior most of the temporal fossa, ensuring exposure comprises at least 6.5 cm behind the temporal tip.
      - Control middle meningeal artery and perform C shaped durotomy.
    - Neocortical resection:
      - Preserve arteries and veins supplying the posterior temporal lobe.
      - Dissection should be carried out in a subpial fashion through the superior temporal gyrus or middle temporal gyrus.
      - Posteriorly the dissection is performed up to the central point or as tailored by functional imaging if the speech area is close by. It should not exceed 3 to 4 cm from the temporal pole along the dominant hemisphere and 5 to 6 cm in the nondominant hemispheres.
      - Use the tentorial edge as guidance to avoid injuring the brainstem and structures within the basal cisterns.
      - Expose the pia of the Sylvian fissure and insula. Enter the temporal horn below the inferior circular sulcus of the insula.
      - The hippocampus and amygdala are identified as glistening white structures.
      - Once the collateral eminence is identified which indirectly indicates the superolateral limit of the lateral ventricular sulcus, the neocorex can be resected en bloc by removing all tissues lateral to it (i.e., superior, middle, inferior, and fusiform gyri).
      - The parahippocampus is aspirated to the level of hippocampal sulcus, which will lead to lateral disconnection of the hippocampus.
    - Hippocampal resection:
      - Use loupe magnification or preferably surgical microscope
      - Removal of the hippocampus is performed by four disconnections
      - Laterally the disconnection is already performed by removing the parahippocampus
      - Anteriorly disconnect through the amygdalohippocampal sulcus, until the choroidal fissure is reached.

## Answers (*continued*)

  - Mesially from the contents of the crural and ambient cisterns through the choroidal fissure. The choroid plexus is protected superiorly, the hippocampal arteries are divided and hippocampus is aspirated.
  - Posteriorly disconnect the tail of hippocampus and the fornix at the level of the tectal plate in the quadrigeminal cistern, 3 cm of hippocampus should be resected.
- The amygdalectomy:
  - Identify the inferior choroidal point. A line is drawn connecting this point to the carotid bifurcation or M1 segment (carotid-choroidal line); resection should not go above this line (▶**Fig. 93.2**).
  - Posterior disconnection is already done through the amygdalohippocampal sulcus.
  - The removal of the remaining uncus is done subpially and guided by visualization of the internal carotid artery, MCA bifurcation, third nerve and posterior cerebral artery to insure complete removal through the transparent inner pia of the uncus.

13. *What is the central point? How does it help you during temporal lobe resection?*
    - The central point is the meeting point between the motor (M) and sensory (S) strips (there may be a connecting sulcus in the hand area of the homunculus at that level referred to as “pli de passage frontopariétal moyen” of Broca[18]).
    - The dotted line indicates extent of resection (▶**Fig. 93.2**).
14. *What is Meyer’s loop and how can it be avoided during temporal lobe surgery?*
    - It represents the part of optic radiation that projects from the relay neurons in the lateral geniculate body (thalamus) forward and lateral.
    - These projections loop on the roof of the temporal horns all the way just beyond them, anterosuperiorly, then backward toward the occipital visual cortex along the calcarine fissure (▶**Fig. 93.3**).[19,20]
    - The loop should be protected during surgery by avoiding the roof of the temporal horns.
    - For selective amygdalohippocampectomy:
      - Transcortical: through the middle temporal gyrus onto the lateral wall of the temporal horn would be a safe pathway.
      - Transsylvian: incisions at the level of the limen insulae, or the adjacent 5 mm of the inferior insular sulcus should be a safe pathway.

**Table 93.1** Various surgical procedures for temporal lobe epilepsy with pros and cons for each

| Procedure | Pros | Cons |
|---|---|---|
| Anterior temporal lobectomy (ATL) | • Significant seizure freedom and improved quality of life | • Risk of neurocognitive disabilities including verbal memory and naming following ATL in the dominant hemisphere[21] |
| Selective amygdalohippocampectomy (SAH) | • Possibly improved neuropsychological outcome[21]<br>• Possibly less risk for verbal memory decline after left SAH compared with left ATL procedure (controversial) | • Slightly worse seizure outcome compared to ATL |
| Transcortical approach | • Technically less demanding.<br>• Minimizes manipulation of Sylvian vessels[22] | • Damage to lateral cortex with risk of development of cortical seizure |
| Transsylvian-approach | • Spares the lateral temporal lobe structures.<br>• Requires minimal temporal lobe retraction[23] | • Risk of vascular injury during Sylvian fissure dissection<br>• Technical complexity and narrow operative corridor require familiarity with microsurgical techniques[23] |
| Subtemporal approach | • Spares the lateral temporal lobe structures[23]<br>• Lower risk for visual field defect | • Increased need for temporal lobe retraction.<br>• Risk of injury to the vein of Labbé[23] |
| Responsive neurostimulation (RNS NeuroPace) | • Option for patients who failed resective surgery or not a candidate<br>• Option for patients with 2 seizure foci<br>• Substantial reductions in seizure frequency<br>• Improvements in measures of quality of life.<br>• Intermittent stimulation so longer battery life and less side effects<br>• No negative effects on mood or depression[24] | • Seizure freedom is rare<br>• Epileptogenic zone localization is required<br>• Need to identify trigger<br>• Possible hardware complications<br>• Need for battery change |
| Bilateral anterior thalamic nucleus stimulation (DBS) | • Option for patients who failed resective surgery or not a candidate<br>• Epileptogenic zone localization is not required.<br>• Reductions of seizure frequency 40% acutely, and 50–69% after several years[25]<br>• Epileptogenic zone localization is not required | • Seizure freedom is rare<br>• The maximal effect seen typically 1–2 years after implantation[25]<br>• Possible hardware complications<br>• Need for battery change<br>• Continuous stimulation so shorter battery life and more side effects |
| Vagal nerve stimulation (VNS) | • Option for patients who failed resective surgery or not a candidate<br>• Epileptogenic zone localization is not required.<br>• A 50% or greater reduction in seizure frequency[26] | • Seizure freedom is rare<br>• The results for VNS are clearly inferior to resective surgery[26]<br>• Possible hardware complications<br>• Need for battery change |

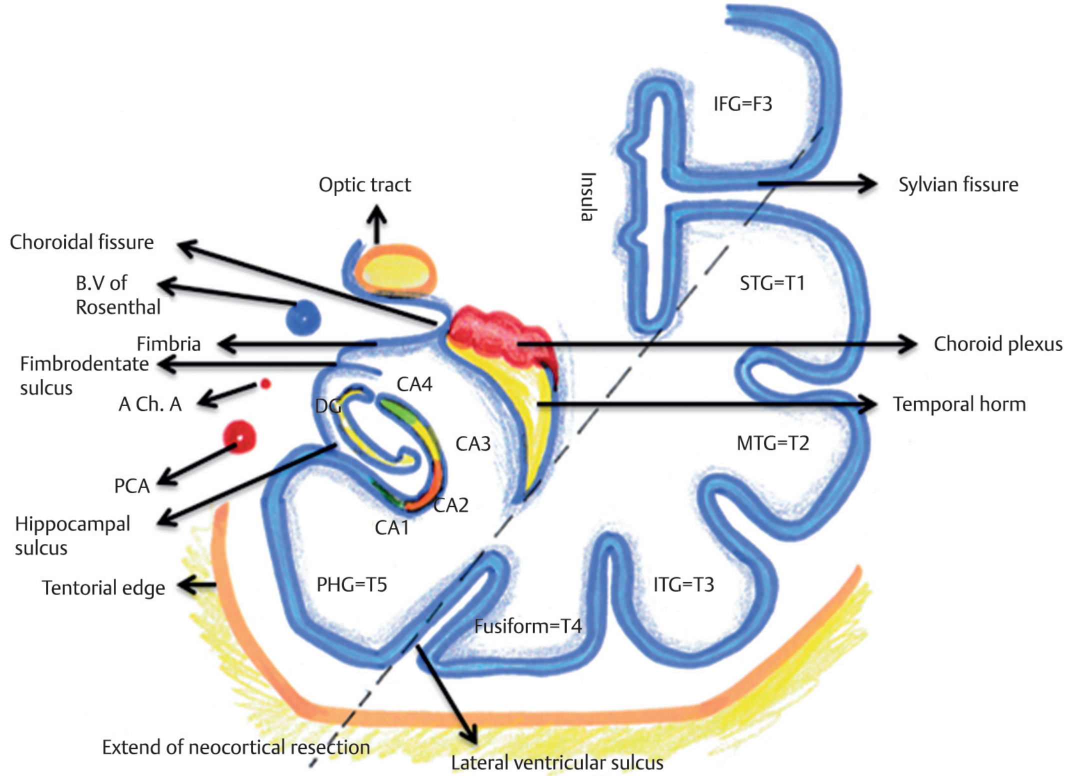
IFG=F3
Optic tract
Insula
Sylvian fissure
Choroidal fissure
B.V of Rosenthal
STG=T1
Fimbria
Choroid plexus
Fimbrodentate sulcus
CA4
A Ch. A
DG
Temporal horm
CA3
MTG=T2
PCA
CA2
CA1
Hippocampal sulcus
Tentorial edge
PHG=T5
ITG=T3
Fusiform=T4
Extend of neocortical resection
Lateral ventricular sulcus

**Fig 93.2** Schematic representation of coronal view of the mesial temporal lobe, Dotted line represents trajectory for the neocortical disconnection. **CA** cornu ammonis, **IFG** (**F3**) inferior frontal gyrus, **STG** (**T1**) superior temporal gyrus, **MTG** (**T2**) middle temporal gyrus, **ITG** (**T3**) inferior temporal gyrus, **PHG** (**T5**) parahipoocampus gyrus, DG dentate gyrus, **PCA** posterior cerebral artery, **A Ch.A** anterior choroidal artery, **B.V. of Rosenthal** basal vein of Rosenthal.

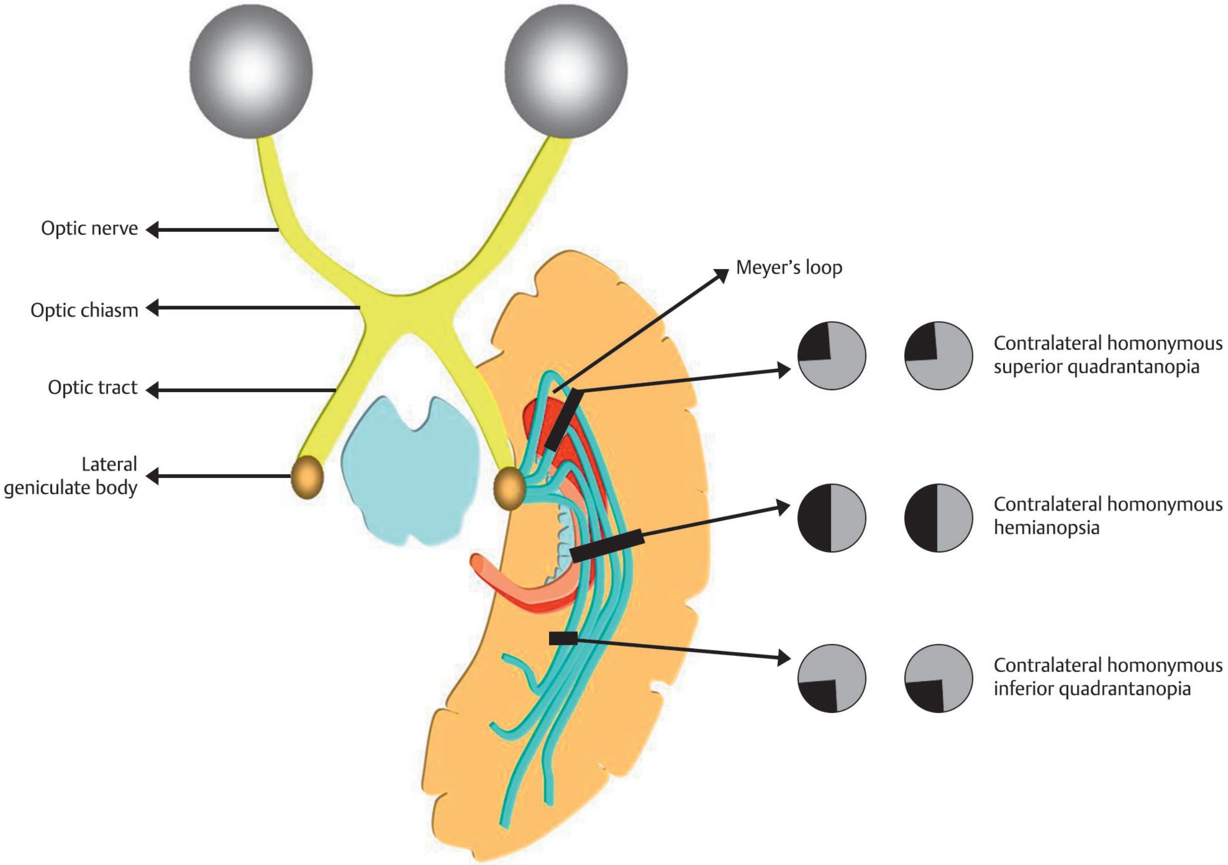
Optic nerve
Optic chiasm
Optic tract
Lateral geniculate body
Meyer's loop
Contralateral homonymous superior quadrantanopia
Contralateral homonymous hemianopsia
Contralateral homonymous inferior quadrantanopia

**Fig. 93.3** Artist's drawing of Meyers's loop and the visual system.

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# Case 94 Hemispherectomy

*Turki Elarjani, Abdulrahman Albakr, Ibrahim Althubaiti, and Salah Baz*

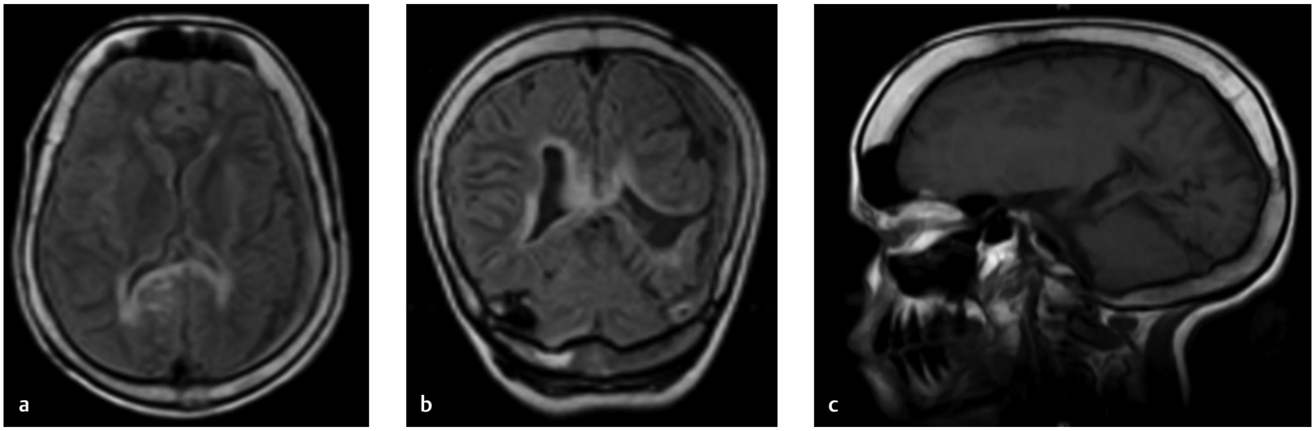

**Fig. 94.1** MRI scan of the brain; T2 fluid-attenuated inversion recovery MRI of the brain axial section (**a**), coronal section (**b**), and sagittal section (**c**).

## Clinical Presentation

- A 22-year-old right-handed woman with known epilepsy since age of 4 years is admitted as an inpatient with medically intractable epilepsy due to recurrent seizure episodes.
- She is on multiple antiepileptic medications, since the age of 4 years.
- The seizure episodes are described to begin with an aura manifesting with numbness and hot sensation in the right hand, followed by impaired consciousness, lip smacking, and eye blinking. She subsequently loses consciousness while her upper limbs are in a fencing posture, her left elbow being extended and her right elbow flexed, terminating in a tonic–clonic episode.
- There is no family history of seizure episodes. She started to walk at the age of 2 years, with normal lower limb and hip development; she is mentally disabled, lives with her parents, and is currently attending a school for special needs.
- On examination, she has a large head circumference, impaired cognition and mentation, spastic dysarthric speech, and diffuse motor spasticity with bilateral positive Babinski sign, yet preserved power in all limbs and approximately normal use of her hand on both sides.

## Questions

1. From the semiology can you localize and lateralize the epileptic focus?
2. What are the investigations of choice that can support your diagnosis?
3. MRI brain has been requested; describe the findings on the image/s (▶**Fig. 94.1**).
4. What is the differential diagnosis based on MRI.
5. Could you mention the types of schizencephaly?
6. How common is schizencephaly associated with epilepsy and what is the percentage of medically intractable seizures?
7. If surgical treatment is an option, what type of surgery is recommended for this patient?
8. What are the indications of hemispherectomy? Please mention pathologies.
9. What are the types of hemispherectomy? Discuss advantages and disadvantages of each.
10. What are the possible complication/s or expected sequalae of hemispherectomy?
11. When is hemispherectomy contraindicated?
12. Is there a role for hemispherectomy in acute cases? Discuss.
13. What is the expected seizure and functional outcome after a hemispherectomy procedure?

## Answers

1. *From the semiology can you localize and lateralize the epileptic focus?*[1]
   Based on the aforementioned auras and ictal presentation:
   - Numbness and hot sensation in the right hand:
     - Lateralized and localized to the contralateral primary somatosensory area which corresponds to the postcentral gyrus (Brodmann area 1–3)
   - Lip smacking:
     - Localized to the mesial temporal lobe, however it does not lateralize to any hemisphere
   - Bilateral eye blinking:
     - Localized to the occipital lobe, however it cannot be lateralized
   - Fencing posture ("figure of 4"), with an extended left elbow and flexed right elbow:
     - Localized to the supplementary motor area (SMA) or prefrontal areas contralateral to the extended elbow side
2. *What are the investigations of choice that can support your diagnosis?*
   - Detailed history taking and physical examination
   - Neuropsychological evaluation
   - Wada test (intracarotid amobarbital procedure in assessing and lateralizing memory and speech in patients preoperatively, mimicking unilateral temporal lobectomy on the site of injection, mainly to evaluate the contralateral temporal lobe's memory and speech functions)
   - EEG; EEG video monitoring if EEG results are uncertain or inconclusive; invasive EEG (epidural, subdural, or parenchymal) is used as a last resort if noninvasive methods (MRI or EEG) are discordant.
   - Intraoperative functional brain mapping
   - MRI of the brain
3. *MRI brain has been requested; describe the findings on the image/s (▶***Fig. 94.1***).*
   - Multifocal areas of significant cortical irregularity and thickening involving the Sylvian region on the left side with associated cerebrospinal fluid (CSF) communication between the lateral ventricles through the temporal lobe into the surface with an associated polymicrogyria.
   - Periventricular hyperintensities are present especially along the occipital regions bilaterally
   - Markedly distorted brain parenchyma
4. *What is the differential diagnosis based on MRI.*
   - Focal cortical dysplasia
   - Porencephaly
   - Grey matter heterotopia
   - Holoprosencephaly
   - Intracranial cyst (such as arachnoid cyst)
5. *Could you mention the types of schizencephaly?*[2]
   - Open lip (separated)
   - Closed lip (fused)
6. *How common is schizencephaly associated with epilepsy and what is the percentage of medically intractable seizures?*[2,3]
   - Thirty-seven to 65% of schizencephaly cases are associated with epilepsy, more common with the open lip type.
   - Nine to 38% of these patients have medically intractable seizures.
7. *If surgical treatment is an option, what type of surgery is recommended for this patient?*
   - Given that she has medically intractable seizures in conjunction with a congenital neural migration disorder (schizencephaly), a functional hemispherectomy is recommended.
8. *What are the indications of hemispherectomy? Please mention pathologies.*[4]
   Hemispherectomy is indicated in cases of intractable epilepsy associated with the following disorders:
   - Acquired disorders such as perinatal cerebral infarction or intracranial hemorrhage, trauma, infection, and hemiconvulsion-hemiplegia-epilepsy syndrome
   - Developmental disorders such as cortical dysplasia, hemimegalencephaly and migration disorders
   - Progressive disorders, such as Rasmussen's encephalitis and Sturge–Weber syndrome
9. *What are the types of hemispherectomy? Discuss advantages and disadvantages of each.*[4–6]
   - Anatomical hemispherectomy
     - Advantages: It is recommended in cases with a small lateral ventricle and dysplastic syndromes.
     - Disadvantages include higher risk of postoperative hydrocephalus.
   - Functional hemispherectomy **(see ▶Fig. 94.2)**
     - Advantages include shorter operative times and less blood losses.
     - Disadvantages: difficulty in evaluating postoperative seizures, in terms of determining the origin of the seizures. Difficulty in determining whether seizures are caused by an incomplete disconnection or are originating from the other cerebral hemisphere.
     - Variants include:
       - Hemispherotomy (trans- and perisylvian and vertical parasagittal techniques)
       - Hemidecortication
10. *What are the possible complication/s or expected sequalae of hemispherectomy?*[4,7]
    - Complications vary among procedures. In addition to standard craniotomy complications, they include:
      - Hydrocephalus
      - Incomplete disconnection and the need for additional epilepsy surgery
      - Mortality rate of 2%

## Answers (*continued*)

- The need for a blood transfusion
- Meningitis
- Postoperative hematoma and stroke
- Adverse pulmonary event

11. *When is hemispherectomy contraindicated?*[8]
    - Hemispherectomy is not performed when the above indications are not met. In addition, the following are considered contraindications:
      - If presurgical evaluation does not demonstrate that all ictal activity is originating from the affected hemisphere.
      - Incomplete transfer of language function is a relative contraindication.
12. *Is there a role for hemispherectomy in acute cases? Discuss.*[9]
    - Hemispherectomy has been used in patients with refractory status epilepticus that does not respond to medical management. It results in improved seizure control and quality of life after surgery.
13. *What is the expected seizure and functional outcome after a hemispherectomy procedure?*[4,10,11,12]
    - In a systematic review, the overall rate of seizure freedom after hemispherectomy was 73.4%. Patients suffering from developmental disorders had lower success rate compared to acquired and progressive disorders.
    - With regards to cognitive outcome, two studies have shown a significant increase of intelligence level after surgery in the pediatrics age group. Similarly, one study reported an increase of IQ by 3 to 18 points in six of eight patients after hemispherectomy.
    - Gradual motor improvement after an initial hemiplegia has been reported in the pediatrics age group.

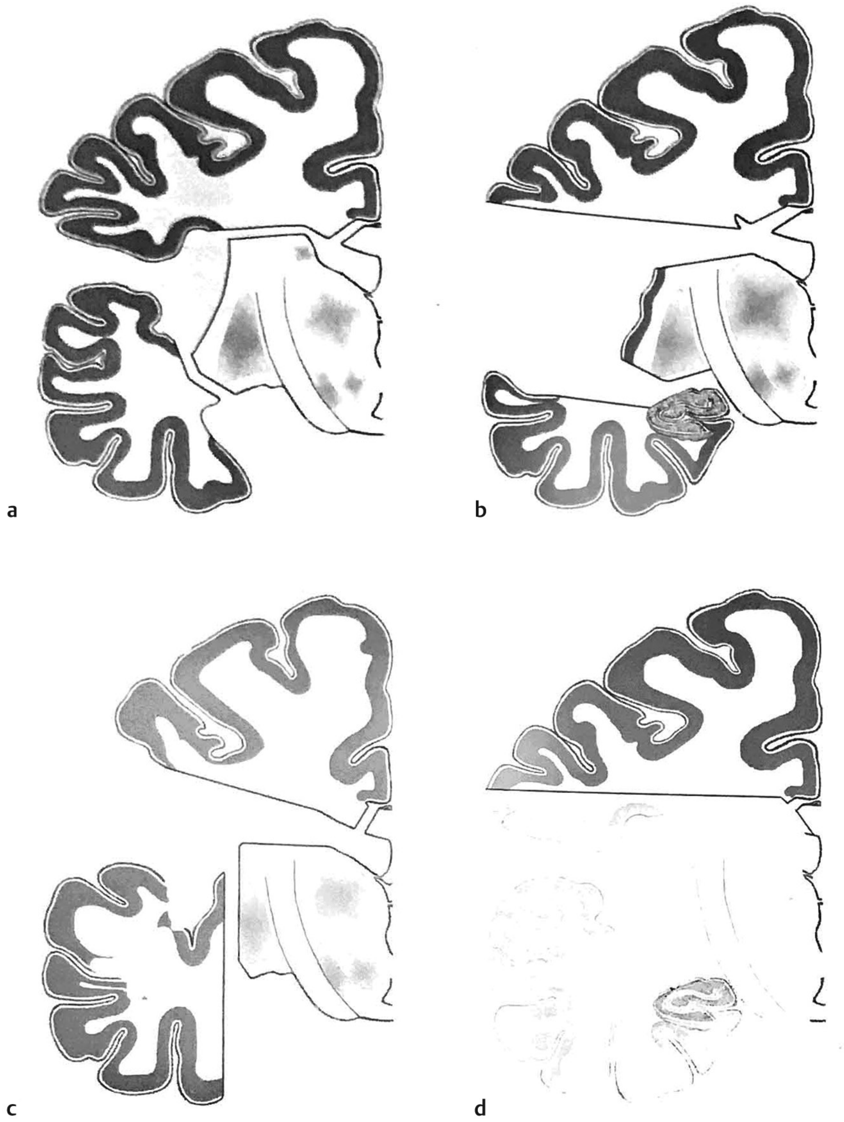

**Fig. 94.2** Artist's rendering of the four types of functional hemispherectomies: (**a**) transsylvian-transventricular approach with temporomesial resection and insular resection. (**b**) Peri-insular window with resection of temporal and frontoparietal operculum and temporomesial disconnection. (**c**) Peri-insular window and temporomesial resection. (**d**) Modified lateral hemispherectomy with removal of temporal lobe, insula, and basal ganglia. (Adapted from Starr P, et al. Neurosurgical Operative Atlas: Functional Neurosurgery. 2nd ed. New York: Thieme.)

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# Case 95 Corpus Callosotomy for Drop Attacks

*Abdulrahman J. Sabbagh, Jeffrey Atkinson, Jean-Pierre Farmer, and José Luis Montes*

## Clinical Presentation

- A 14-year-old right-handed girl was diagnosed with multifocal epilepsy since the age of 4 years.
- Her epilepsy has become progressive with time and has been intractable for the past 3 years.
- Her seizures are described as staring events that occur 2 to 4 times a day and atonic drop attack that occur once or twice daily despite compliance with triple therapy.
- She is on three antiepileptic medications, without which she has frequent generalized tonic–clonic seizures.
- On examination she is somewhat cognitively subnormal. She has multiple scalp scars of different ages from repeated falls.

## Questions

1. How would you investigate this case?
   MRI of the brain was essentially normal. Electroencephalography (EEG) showed bilateral multifocal epilepsy.
2. What are the surgical options?
3. What would you tell the parents regarding expected seizure outcome after surgery?
4. What are the predictors of a better outcome?

   You operate on her and perform an anterior two-thirds corpus callosotomy. Postoperatively she is well and awake but would not interact or speak for the first few days. She returns to her normal self by the end of the week.
5. The parents were concerned; what do you tell them happened?
6. What are the possible complications related to corpus callosotomy?
7. What are the indications for corpus callosotomy?
8. Describe the parts of the corpus callosum.
9. Compare the callosotomy procedure with vagal nerve stimulation.
10. What are the approaches for corpus callosotomy?

## Answers

1. *How would you investigate this case?*
   - Investigations include the following:
     - Drug levels to check compliance
     - Single photon emission CT (SPECT) scans
       - Ictal mode
       - Interictal mode
     - MRI brain
     - Video EEG telemetry
2. *What are the surgical options?*
   - Surgical options for intractable multifocal epilepsy include[1–3]:
     - Corpus callosotomy
       - Anterior two-thirds callosotomy
       - Complete callosotomy (not recommended for almost normal children)
     - Vagal nerve stimulation
3. *What would you tell the parents regarding expected seizure outcome after surgery?*
   - This surgery is palliative.
   - It is more effective in treating atonic or drop attacks compared with other epilepsy types.[1,2]
   - A meta-analysis/systematic review of the literature showed that the long-term seizure outcome is 35% of patients with callosotomy become free of most disabling seizures.[4]
4. *What are the predictors of a better outcome?*
   - Factors predicting better chance of improvement include the following:
     - Lateralization
     - Frontal origin of seizures
     - Extent of corpus callosum resection[1,5,6]
5. *The parents were concerned; what do you tell them happened?*
   - She has developed the expected postcallosotomy transient mutism.[7]
6. *What are the possible complications related to corpus callosotomy?*
   - Postoperative complications are as follows:
     - Mortality risk is less than 1% and morbidity rates are 6 to 30%.[6,8,9]
     - The callosal syndrome includes[8,10,11]:
       - Inability to name objects presented briefly to the left visual hemifield
       - Left hemialexia
       - Left hemianomia
       - Difficulty imitating the hidden other hand

## ■ Answers (*continued*)

        - Unilateral tactile anomia
        - Unilateral left agraphia
        - Right-hand constructional apraxia (inability to copy a complex design with the right hand, but ability to outperform this by using the left hand)
    - Interhemispheric retraction
        - Supplementary motor area injury
        - Cingulate gyrus injury
    - Vascular compromise or injury to the following:
        - Superior sagittal sinus hemorrhage or occlusion
        - Pericallosal and supramarginal artery injury
    - Disconnection syndrome[7]

7. *What are the indications for corpus callosotomy?*
    - Callosotomy is a palliative treatment aimed at seizure reduction rather than seizure cure.
    - It is indicated for intractable multifocal epilepsy that is not amenable to resection of an epileptic focus and that is associated with drop attacks.[1,2]
    - Such examples include patients with severe Lennox–Gastaut syndrome.
8. *Describe the parts of the corpus callosum.*
    - Parts of the corpus callosum are described below (▶ **Fig. 95.1**):
        - Rostrum
        - Genu
        - Body
        - Isthmus
        - Splenium
        - Forceps minor and major
    - ▶ **Fig. 95.1** also shows the approximate locations of connecting fibers of major cortical brain regions.[12]
9. *Compare the callosotomy procedure with vagal nerve stimulation.*
    - Better seizure control can be achieved by callosotomy, especially drop attacks.[3]
    - Vagal nerve stimulation is less invasive.
    - Vagal nerve stimulation is reversible unlike callosotomy.
    - Vagal nerve stimulation has less morbidity.
    - Vagal nerve stimulation requires battery changes and a closer follow-up.[3]
10. *What are the approaches for corpus callosotomy?*
    - Standard anterior two-thirds callosotomy[1] or complete callosotomy[2]
        - Supine position
            - Slight flexion with placement of the midline of the cranium at a right angle to the floor
            - Single or double skin openings (anterior and posterior)
            - Interhemispheric approach by entering the cranium at the nondominant hemisphere and/or the side with the least crossing superior sagittal sinus tributaries obstructing the way (neuronavigation is very helpful in outlining those vessels preoperatively).[13]
        - Lateral position (Olivier technique[1])
            - Letting the side of entry inferior, so the brain will sag with gravity to help open the interhemispheric fissure without retraction (▶ **Fig. 95.2**)
    - Two-stage approach[13]
        - Starting with the two-thirds callosotomy approach and planning for the remaining one-third at a second stage if seizures need better control despite improvement
    - Other techniques
        - Endoscopic approach[14]
        - Gamma knife or other forms of radiosurgery[15]

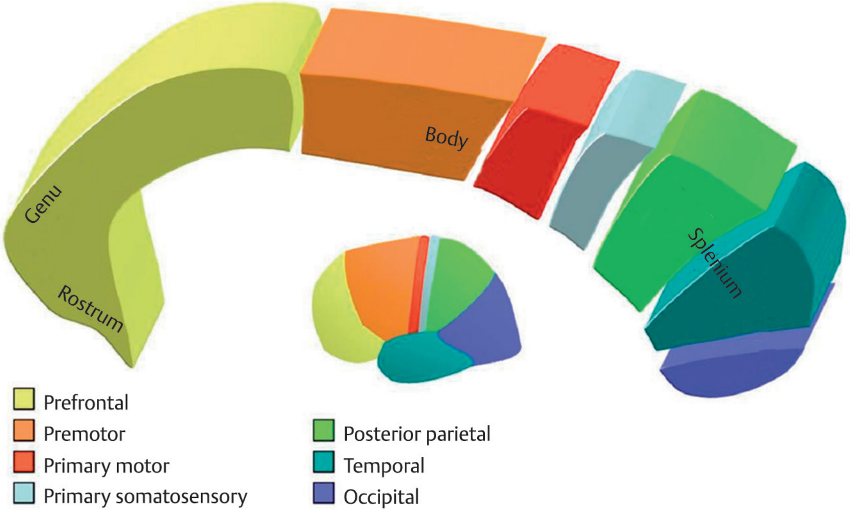

**Fig. 95.1** Parts of the corpus callosum. The approximate locations of connecting fibers of major cortical brain regions are highlighted in the smaller unlabeled image.

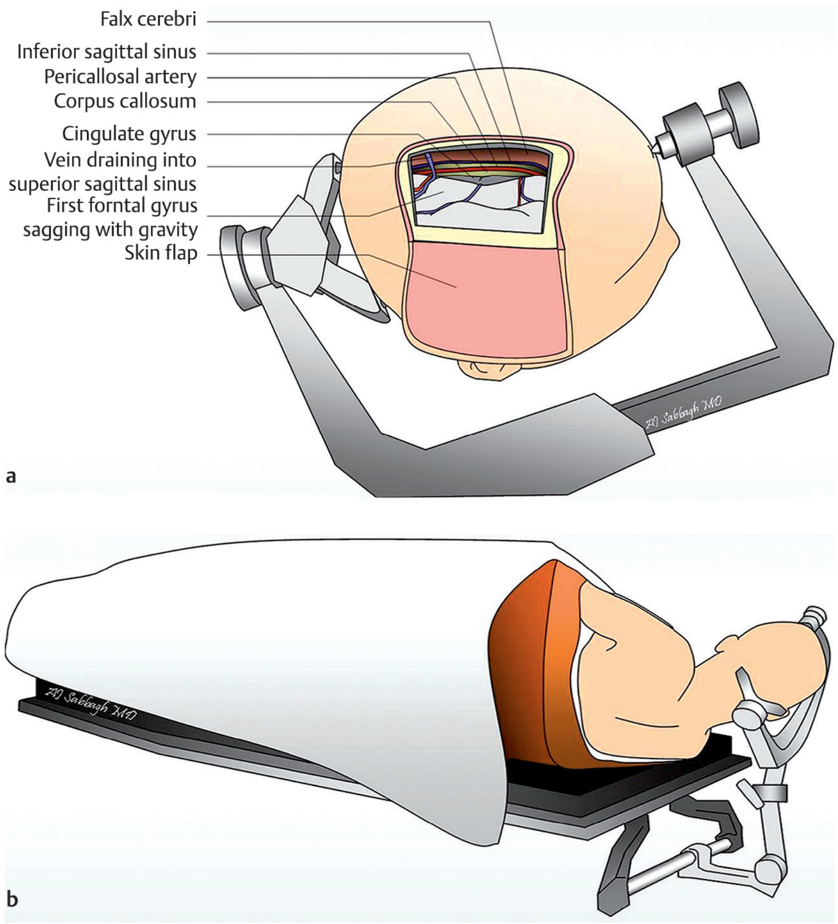

**Fig. 95.2** Corpus callosotomy via lateral positioning. **(a)** Shows head positioning with respect to the floor, bone flap opening, and exposure. Note that the side of entry is inferior so the brain sags with gravity to help open the interhemispheric fissure without the need for retraction. **(b)** Patient positioning on the bed with Mayfield three-point fixation head holder.

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# Case 96 Vagal Nerve Stimulator

*Nazer Qureshi and Remi Nader*

## Clinical Presentation

- A 16-year-old boy with cerebral palsy and a long history of seizures is referred to you for a vagal nerve stimulator (VNS) placement.
- The mother wishes the generator to be placed on the right side because of flexion contractures in the left upper extremity.

## Questions

1. What is the mechanism of action of VNS?
2. What is the United States Food and Drug Administration (FDA)-approved indication for VNS in the treatment of epilepsy?
3. How do you identify the vagal nerve in the neck for implantation?
4. Where on the vagal nerve should the lead wires be ideally placed, and is there any concern regarding VNS placement on the right side?
5. What are the reported results for VNS use in epilepsy?
6. What are the usual initial VNS settings for use?
7. What are the usual side effects of VNS placement?
8. The patient returns to you a month later with pus draining out from the battery site. What are your options?
9. Antibiotics and your debridement cleared the infection. What are you going to do next?
10. A few weeks after reimplantation of the battery at the new site, the patient presents with infection along the leads extending into the neck. What are your options now?
11. Nine months after all the infection is cleared, the patient is referred to you once more for reimplantation of VNS. What are your options now?
12. Other than the treatment of seizures, is there any other indication for VNS placement?

## Answers

1. *What is the mechanism of action of VNS?*
   - The precise mechanism of action of VNS is unknown.
   - It has been suggested that VNS has action on various regions of the brain including the locus ceruleus, amygdala, hippocampus, and contralateral somatosensory cortex.
   - Another proposed mechanism of action is the possibility that VNS increases γ-aminobutyric acid (GABA) and glycine levels in the brain.
   - Ben-Menachem et al[1] measured amino acid and neurotransmitter metabolite concentrations in cerebrospinal fluid (CSF) samples of patients on clinical trials of VNS before and 3 months after VNS placement.
   - Their results showed an increase in GABA level among patients that failed to respond to VNS stimulation, those with a lower setting of stimulation, as well as patients who had undergone a long-term VNS stimulation.
   - Yet others have indicated that VNS decreases cortical epileptiform activity directly and possibly also affects blood flow through different regions of the brain.[2,3]
2. *What is the FDA-approved indication for VNS in the treatment of epilepsy?*
   - In 1997, the FDA approved implantation of VNS as an adjunctive therapeutic modality in reducing the frequency of seizures in adults and adolescents over 12 years of age with partial-onset seizure that are refractory to antiepileptic medications.[4]
   - Although the FDA indication for VNS excludes other types of epilepsies, most epileptologists and neurosurgeons believe the indications for placement of VNS to be more widespread.[5,6]
3. *How do you identify the vagal nerve in the neck for implantation?*
   - The vagal nerve is in the posterior part of carotid sheath between the carotid artery and internal jugular vein.
   - See ▸**Fig. 96.1** describing electrode placement.
   - Caution is advised to differentiate the vagus nerve from the phrenic nerve, which traverses in the anterior part of the sheath.
4. *Where on the vagal nerve should the lead wires be ideally placed, and is there any concern regarding VNS placement on the right side?*

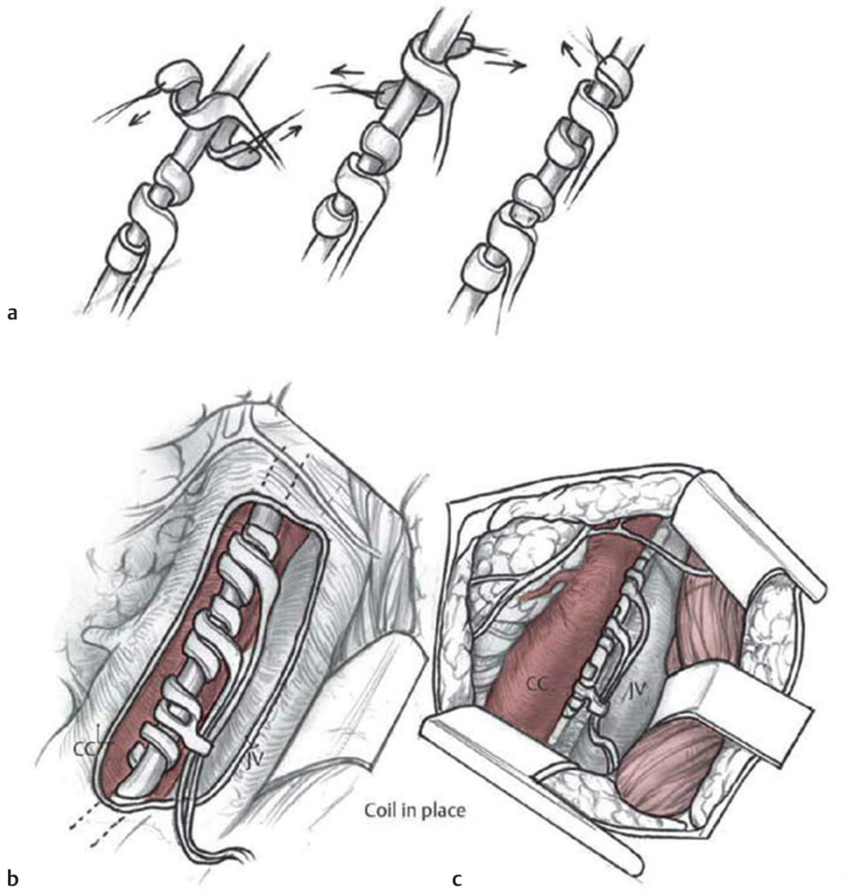

**Fig. 96.1** Implantation of the vagal nerve leads. Placement is performed by initially grasping the embedded sutures. The loops of the coil are gently wrapped around the nerve. (**a**) The electrode tension strings are looped around the vagus nerve. (**b**) The electrode naturally coils around the nerve making it easier to position. (**c**) Final electrode configuration and final position in relation to surrounding structures. JV, jugular vein; CC, common carotid artery. (Adapted with permission from Nader et. al. Neurosurgery Tricks of the Trade—Cranial, Thieme: New York; 2014.)

## Answers (*continued*)

- The right vagal nerve innervates the sinoatrial (SA) node more than 60%, whereas the left vagal nerve mostly innervates the atrioventricular (AV) node of the heart.
- Placement of the leads on the right vagal nerve is contraindicated, as the stimulation will result in asystole.
- The risk of bradycardia exists even when the electrodes are placed on the left vagal nerve.
- What the mother wished was for the battery pocket to be placed on the right side secondary to flexion contractures of the left upper extremity. This can be done and should be considered given the contractures.[7]
- The ideal place for implantation of leads on vagus nerve is in the middle part of the neck proximal to the branches of the cardiac plexus.
- The three lead wires are placed from proximal to distal along the nerve.

5. *What are the reported results for VNS use in epilepsy?*
    - In the Vagus Nerve Stimulation Study Group E05 trial, the median reduction in seizure frequency at 12 months after completion of the initial double-blind study was 45%.[8]
    - Overall, 35% of the patients had a reduction of at least 50% in seizures; 20% of the patients demonstrated a 75% reduction in their seizure frequency.[8]
    - Similar results have also been reported in the XE5 trial.[9]
    - A 12-year retrospective review of the effectiveness of VNS in 48 patients with intractable partial epilepsy reported a mean decrease in seizure frequency by 26% after 1 year, 30% after 5 years, and 52% after 12 years.[10]
6. *What are the usual initial VNS settings for use?*
    - Although some centers initiate stimulation the day after implantation, usually the generator is kept turned off and an increase in output is advanced by the neurologists after a 2-week postoperative period.
    - Typically, the output is adjusted to tolerance, using a 30-Hz signal frequency, with a 500-microsecond pulse width for 30 seconds of “on” time and 5 minutes of “off” time.
    - This is not standard and multiple variations exist depending on the clinical situation.
7. *What are the usual side effects of VNS placement?*
    - The most common side effects of VNS are cough, hoarseness, and throat pain.
    - Unlike antiepileptic drugs, VNS has not been associated with adverse effects such as depression, fatigue, confusion, or cognitive impairment, etc.

## Answers (*continued*)

8. *The patient returns to you a month later with pus draining out from the battery site. What are your options?*
   - Infection at the site of battery would necessitate removal of the battery and treatment with debridement and antibiotics.[11]
   - The lead wire could be left in place and moved away from the infected site.
9. *Antibiotics and your debridement cleared the infection. What are you going to do next?*
   - Once the infection is cleared, a new battery should be implanted at another site in the anterior chest wall and reconnected to the lead wires.
10. *A few weeks after reimplantation of the battery at the new site, the patient presents with infection along the leads extending into the neck. What are your options now?*
    - If there is suspicion of infection along the lead wires, then the neck incision should be opened and explored.[12]
    - The wires should be cut close to the vagus nerve's lead implantation site and the leads should be left in situ.
    - Attempting to remove the leads from the vagus nerve will result in injury to the nerve.
11. *Nine months after all the infection is cleared, the patient is referred to you once more for reimplantation of VNS. What are your options now?*
    - The vagus nerve can be explored proximally in the neck and if it is possible to implant another set of leads on the vagus nerve, it should be done proximal to its cardiac branches.
12. *Other than the treatment of seizures, is there any other indication for VNS placement?*
    - Other indications for use of VNS include refractory depression, and research is being conducted for its use in the treatment of such varied diseases as anxiety disorders, Alzheimer's disease, migraines, and fibromyalgia.

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# Case 97 Idiopathic Intracranial Hypertension

*Charles B. Agbi and Mohammad Misfer Alshardan*

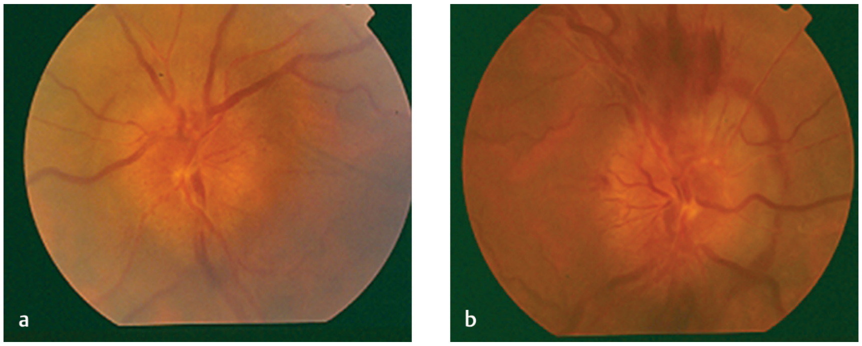

**Fig. 97.1** Fundoscopic examination findings of the left (**a**) and right (**b**) eyes.

## Clinical Presentation

- A 23-year-old woman was referred to the clinic with a history of severe headaches, visual blurring, and phonophobia. The symptoms had progressively worsened, and her vision had become more blurred especially over the last month. The headaches were severe and continuous, and localized to the frontal and temporal region.
- Clinical examination showed a body mass index (BMI) of 38 (height 1.67 m, weight 106 kg). Her visual acuity was 20/150 OD, 20/150 OS.
- Fundoscopic examination is depicted in ▶**Fig. 97.1**
- The MRI scan shows:

1. Bilateral flattening of the posterior aspect of the sclera (▶**Fig. 97.2**)
2. Partial “empty sella” with flattening of the hypophysis (▶**Fig. 97.3**)
3. Mild focal narrowing of the distal transverse sinuses, worse on the right side

## Questions

1. What is the most likely diagnosis?
2. Describe the funduscopic findings.
3. What do the MRI findings indicate (▶**Fig. 97.2** and ▶**Fig. 97.3**)?
4. What other imaging findings are seen in this condition?
5. What additional confirmatory test would you recommend?
6. What epidemiologic factors are associated with this condition?
7. Discuss theories of the pathophysiology of idiopathic intracranial hypertension (IIH).
8. What is the natural history of this condition?
9. What treatment options are currently available for this condition?
10. Discuss the long-term management.

## ■ Answers

1. *What is the most likely diagnosis?*
   - The most likely diagnosis is IIH.
   - This condition often presents, as in this case with headaches and visual symptoms.[1,2]
     - The headaches are usually severe and often localized to the frontal, retro-ocular, and temporal regions.
     - Common visual symptoms include blurred vision and loss of peripheral vision. Other visual symptoms include diplopia, which occurs in one-third of the patients, and is thought to be due to sixth nerve palsy due to the high intracranial pressure (ICP; "false localizing sign").
2. *Describe the funduscopic findings.*
   - The funduscopic pictures show bilateral florid papilledema. This finding is consistently present in IIH, and along with elevated cerebrospinal fluid (CSF) pressure, is one of the findings required for diagnosis.[1,2]
   - The papilledema is probably the reason for most of the visual symptoms. Objectively, visual field assessment may reveal constriction of the visual fields, loss of color vision, and decreased visual acuity.
   - If untreated, consecutive optic atrophy with complete visual loss may occur. The objective findings are usually bilateral, although they may occasionally be asymmetrical.
3. *What do the MRI findings indicate (*▶ **Fig. 97.2** *and* ▶ **Fig. 97.3***)?*
   - The MRI shows flattening of the posterior aspect of the sclera and a partial "empty sella."
   - These findings are indicative of increased ICP due to increase CSF pressure.
4. *What other imaging findings are seen in this condition?*
   - MRI is often helpful in the diagnosis of IIH.[3–5] ▶ **Fig. 97.3**, ▶ **Fig. 97.4**, and ▶ **Fig. 97.5** show some of the other common radiological findings in IIH. These findings include:
     - Protrusion of the optic nerve heads into the vitreous
     - Postgadolinium enhancement of the optic nerve heads and tortuosity of the nerve sheath complex
   - Venous sinus stenosis, especially stenosis of the transverse sinus, is often found in this condition[3–7] (▶ **Fig. 97.5**). The stenosis is sometimes the result of sinus thrombosis, which may explain the association between this condition and oral contraceptive use as well as in other hypercoagulable states.[3]
   - One of the authors' IIH cases had repeated episodes of venous sinus thrombosis with one severe episode resulting in cerebral deep venous thrombosis. The patient was discovered to have polycythemia vera.
   - Arteriovenous malformations (AVMs) with superficial venous sinus drainage associated with IIH has been reported.[8] One of the authors' patients (▶ **Fig. 97.5**) was discovered to have an AVM with drainage into the distal superior sagittal sinus. In this case, the IIH resolved, following successful obliteration of the AVM with radiosurgery. The elevated venous pressure is thought to be responsible for the increased CSF pressure in these cases.
5. *What additional confirmatory test would you recommend?*
   - A lumbar puncture (spinal tap) is required for the confirmation of the diagnosis of IIH.[1–3]
   - Typically, pressures > 25 cm $H_2O$ are confirmatory.
   - The modified Dandy criteria[2] have been proposed for the diagnosis of IIH, and are helpful in patients with CSF opening pressure between 20 and 25 cm $H_2O$.
   - A lumbar puncture also offers the opportunity to relieve symptoms by removing CSF. Such CSF removal can prevent rapid deterioration of visual symptoms, while more definitive treatment is being arranged. Prolonged symptom relief has been observed following CSF removal during lumbar puncture.
6. *What epidemiologic factors are associated with this condition?*
   - By far the most important epidemiological factors in IIH are high BMI[3] and female gender.[3]
     - Most patients are obese (BMI > 30) or overweight (BMI 25–30). Even in nonobese women, the incidence of IIH is significantly higher in cases where there is a recent weight gain. There have also been reports of the resolution of IIH following successful treatment of obesity.[3,9]
     - In females, in addition to the BMI, there is a fairly strong association between IIH and current or recent use of oral contraceptives.
   - IIH is rare in men who constitute approximately 10% of patients. They are often older and not obese, and potential causative findings such as venous sinus stenosis and/or thrombosis are more common.
   - Conversely, IIH is very rare in the pediatric age group. Obstructive sleep apnea (OSA) is another condition sometimes associated with IIH.[22] In addition to oral contraceptives, IIH has been associated with certain medications, of which the tetracyclines have been the most supported by evidence.[10,11] Others include vitamin A, lithium, cyclosporine, human growth hormone, and tamoxifen.[11]
7. *Discuss theories of the pathophysiology of idiopathic intracranial hypertension (IIH).*
   - The exact pathophysiology of IIH is poorly understood.
   - Traditionally, three pathophysiological mechanisms have been proposed:

## Answers (*continued*)

    - Overproduction of CSF
    - Poor reabsorption of CSF
    - CSF outflow obstruction
  - None of these mechanisms completely explain the association with obesity or gender.
  - However, it is likely that the pathophysiology is multifactorial.[3,6,8]
  - The response to agents, such as acetazolamide, which decrease CSF production suggests that excessive CSF production may play a part,[12] but no strong evidence exists to support abnormality of reabsorption in cases of IIH.
  - The recent identification of cerebral venous outlet obstruction (venous sinus stenosis) in several cases of IIH has increased interest in this as a possible pathophysiological mechanism.
  - The role of vitamin A metabolism in the pathophysiology of IIH has been studied extensively.[12] The studies have yielded suggestive but inconclusive results. However, the association of abnormal vitamin A metabolism with some types of obesity has led to increased interest in this area of inquiry.[9]
  - A hypercoagulable state may play a role in some cases, if, as in the case referred to above, they lead to venous sinus thrombosis.

8. *What is the natural history of this condition?*
   - There are no data on the natural history of untreated cases.
   - However, observations in medically treated cases suggest that while the symptoms may resolve in some cases, many require long-term treatment, and in fact, the symptoms may worsen with time.
   - Even patients who respond to surgical treatment by shunting often relapse if the shunts fail, even after long periods. It is likely, therefore, that for most patients, the condition pursues a chronic course.
9. *What treatment options are currently available for this condition?*
   - Treatment options include general measures to address risk factors such as obesity, and medical treatment and surgical techniques aimed at reducing symptoms and especially at preventing permanent visual loss.
   - Successful weight loss through dietary and lifestyle changes, and occasionally bariatric surgery has resulted in resolution or better control of the symptoms in IIH patients.[15] OSA should also be treated when present, although it is not clear whether such treatment results in control of the IIH.[22]
   - Medical management with acetazolamide is often successful in relieving the symptoms and reducing the CSF pressure, and it is often the first line of treatment for most patients. Other carbonic anhydrase inhibitors, such as topiramate are also effective, particularly in patients with serious side effects from acetazolamide. Loop diuretics (furosemide, etc.), have also been tried.
   - Therapeutic lumbar puncture, with removal of 30 to 50 mL of CSF results in rapid (and sometimes prolonged) relief of symptoms, and can be used in situations of rapid visual deterioration to temporize until definitive treatment can be arranged.
   - There are currently three surgical options:
     - Optic nerve sheath fenestration (ONSF)
     - CSF diversion
     - Venous sinus stenting
   - ONSF is indicated in patients with papilledema but with nondebilitating headaches. It is safe and relatively effective.[13,14] As with other surgical procedures, failure rates are high, and repeat surgery carries significant risks of morbidity and is therefore rarely advised.[14]
   - CSF diversion is achieved either by ventriculoperitoneal (VP) or lumboperitoneal (LP) shunting. Both techniques have been shown to be effective in controlling the symptoms and signs of IIH.[15–18] Both are subject to high failure rates and also high rates of short- and long-term complications.
     - VP shunts are difficult to place in the presence of slit-like ventricles, and where possible, navigation techniques should be used to prevent malposition or placement-related complications.[17] Even when properly placed, failure rates may be higher than for LP shunts, possibly because of the collapse of the ventricle around the catheter.
     - LP shunts are often complicated with morbidity from low-pressure headaches and acquired tonsillar descent. These problems can be somewhat largely avoided with the use of programmable valves. We have successfully used LP shunts in patients where properly placed and patent VP shunts failed to maintain CSF pressure reduction.
   - Recently, endovascular stenting for venous (usually transverse) sinus stenosis has been shown to result in decreased CSF pressure and symptom relief in IIH patients.[6,7] This technique should be considered in cases where there is clearly demonstrated stenosis and a transverse sinus pressure gradient > 8 to 10 mm Hg. Complications of this technique include sinus thrombosis and occlusion, stent migration, and sinus perforation. More data are needed to determine the proper role of this technique in the management of IIH.
10. *Discuss the long-term management.*
    - The majority of cases pursue a chronic course and therefore require regular long-term clinical and especially neuro-ophthalmological follow-up to monitor the visual function.
    - Patients treated with CSF shunting or venous sinus stenting also require long-term systematic follow-up.

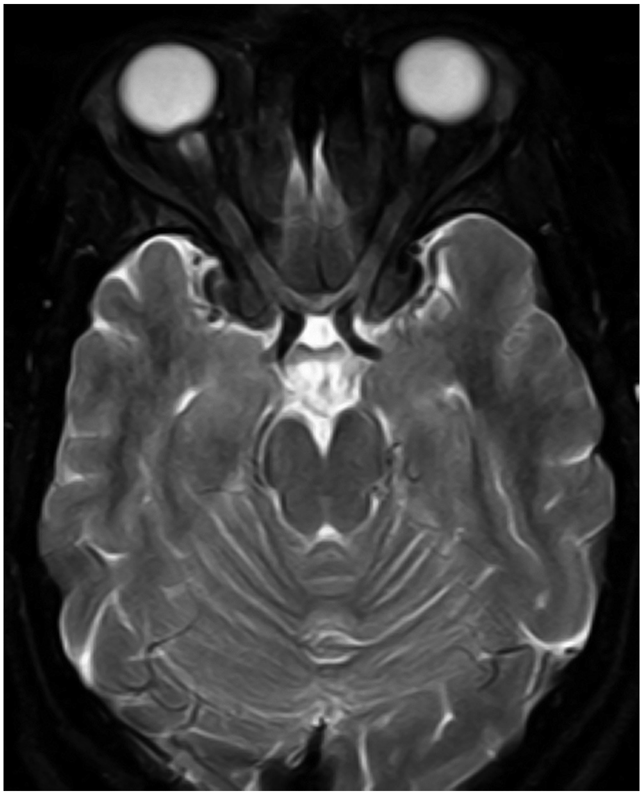

**Fig. 97.2** Axial T2-weighted MR image at the level of the optic nerve and chiasm. Note the bilateral flattening of the posterior aspect of the sclera.

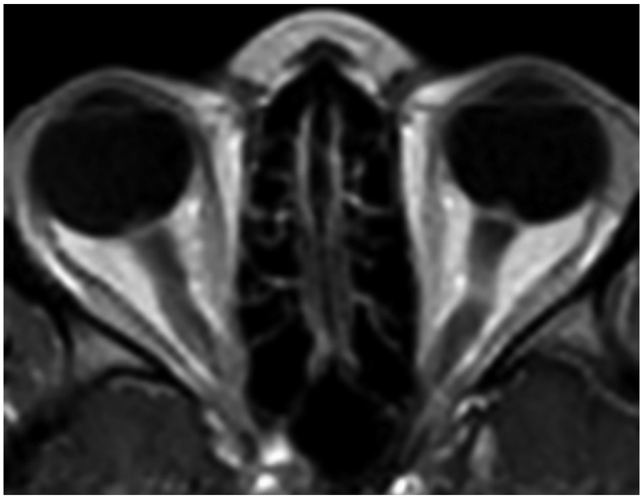

**Fig. 97.4** Axial T2-weighted MR image at the level of the optic nerve of a different idiopathic intracranial hypertension patient showing protrusion of the optic nerve heads into the vitreous.

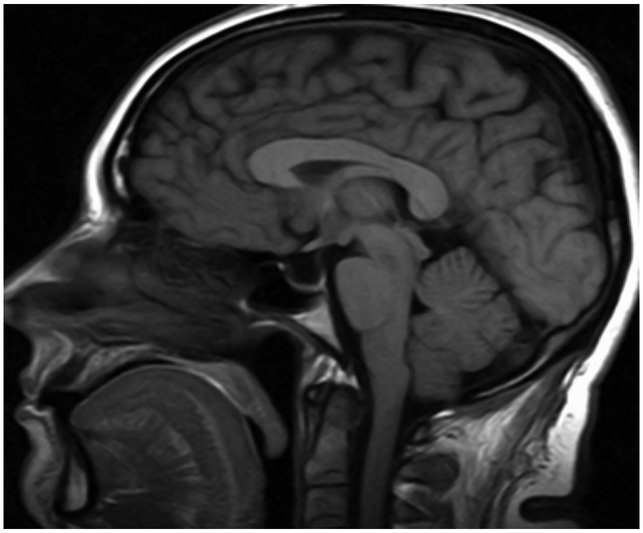

**Fig. 97.3** Midsagittal T1-weighted MR image showing partial "empty sella" with flattening of the hypophysis.

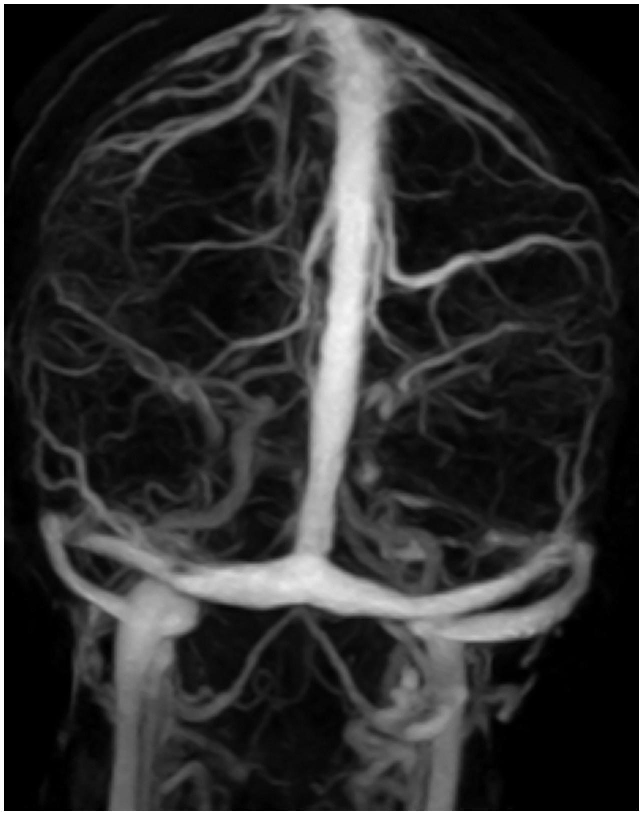

**Fig. 97.5** MR venography reconstructed images of another idiopathic intracranial hypertension patient showing stenosis of the transverse sinus.

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# Case 98 Normal Pressure Hydrocephalus

*Remi Nader*

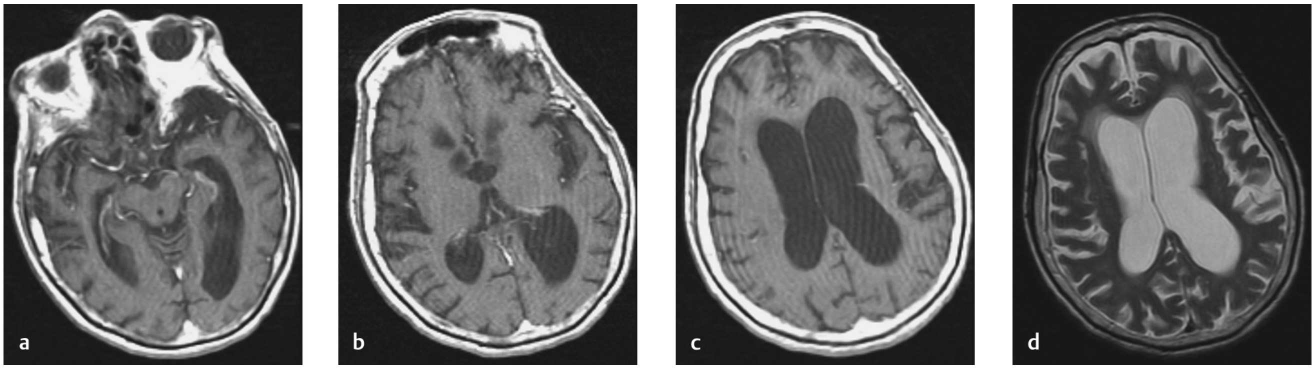

**Fig. 98.1** (**a–c**) Sequential axial T1-weighted MR images of the brain with infusion of intravenous contrast. (**d**) A T2-weighted axial image at the level of the lateral ventricles is shown.

## Clinical Presentation

- A 64-year-old woman with dementia and a history of previous head injury ~30 years ago presents with progressive deterioration in her mental status with further dementia as well as incontinence over the past year.
- Over the past 2 months, she has been having some increased drowsiness as well as decreased appetite and some weight loss. She is becoming more agitated.
- She has had some decreased vocalization in the past year.
- The patient has been nonambulatory for ~6 months.
- The remainder of her neurologic examination is within normal limits.
- MRI is obtained and pertinent images are shown in ▶**Fig. 98.1**.

## Questions

1. Interpret the MRI.
2. Provide a differential diagnosis and the most likely diagnosis.
3. What are the main clinical criteria for the most likely diagnosis?
4. What is Evans' ratio?
5. What other conditions predispose to this diagnosis?
6. What diagnostic studies can confirm this diagnosis?
7. What are the limitations of these studies?
8. What treatment measures are available for this condition?
9. What are the main complications of these treatment measures?
10. Name some prognostic factors for the treatment of this condition.

## Answers

1. *Interpret the MRI.*
   - MRI shows diffuse brain atrophy and ventriculomegaly.
   - The degree of ventriculomegaly appears to be disproportionately elevated compared with the amount of atrophy.
   - There appears to be some transependymal cerebrospinal fluid (CSF) transudation in the form of periventricular high signal on T2-weighted images.
2. *Provide a differential diagnosis and the most likely diagnosis.*
   - Normal pressure hydrocephalus (NPH)—most likely diagnosis
   - Differential diagnosis of dementia includes (mnemonic is "CITTEN DIVA," more common conditions are in italics)[1,2]:
     - Congenital/developmental: Huntington's disease
     - Infectious: syphilis, human immunodeficiency virus (HIV), meningitis, herpes encephalitis

## Answers (*continued*)

  - Traumatic: *posttraumatic dementia*, concussion, chronic subdural hematoma, hypoxia
  - Tumor: metastatic disease, carcinomatosis
  - Endocrine: Addison's disease, Cushing's syndrome, diabetes mellitus, thyroid and parathyroid disease, renal failure
  - Neurologic: *Alzheimer's disease*, Parkinson's disease, Pick's dementia
  - Drugs/medications: alcohol induced, vitamin B12 or folate deficiency, pellagra, vitamin $B_1$ deficiency
  - Inflammatory: multiple sclerosis, prion disease
  - Vascular: diffuse small vessel disease, stroke
  - Acquired and other: depression, psychosis

3. *What are the main clinical criteria for the most likely diagnosis?*
   - The Adams triad includes[3]:
     - Ataxia: precedes other symptoms, wide based, "glued to the floor," difficult initiation of gait
     - Dementia: memory deficit, bradyphrenia, bradykinesia
     - Urine incontinence
   - Other criteria include male sex, age > 60 years, communicating hydrocephalus, normal pressure on lumbar puncture (LP)
4. *What is Evans' ratio?*
   - The ratio of the maximum width of the frontal horns to the maximum width of the inner table of the cranial vault[4]
   - If the ratio is greater than 0.3, then there is a greater likelihood of hydrocephalus.[2]
5. *What other conditions predispose to this diagnosis?*
   - Postsubarachnoid hemorrhage
   - Posttrauma
   - Postmeningitis
   - After posterior fossa surgery
   - Tumor, carcinomatous meningitis
   - Alzheimer's disease
   - Aqueductal stenosis
6. *What diagnostic studies can confirm this diagnosis?*
   - Diagnostic studies are outlined below[2,3,5]:
     - CSF "tap" test by performing an LP with removal of 40 to 50 cc of CSF followed by assessment of improvement of cognitive abilities
     - Serial LP
     - Continuous intracranial pressure (ICP) monitoring
     - Lumbar drain placement
     - Radionucleotide cisternography
7. *What are the limitations of these studies?*
   - Limitations of the studies include the following[2,3]:
     - CSF "tap" test has a poor sensitivity (26–62%).[3]
     - LP: An opening pressure (OP) greater than 10 (but less than 18 mm $H_2O$) is associated with a higher response rate to shunting.
     - Continuous ICP monitoring: Normal OP, but pressure peaks greater than 270 mm $H_2O$ or recurrent B waves are predictors of better prognosis with shunting.
     - Radionucleotide cisternography: Persistence of ventricular activity in a late scan (after 48–72 hours) is associated with a 75% chance of improving with shunting (this is also the case if the ratio of ventricular to total intracranial activity (V/T) is greater than 32%).
8. *What treatment measures are available for this condition?*
   - Ventriculoperitoneal shunt
     - Medium-pressure valve shunt
     - Programmable shunt
   - Other shunt types: ventriculopleural or ventriculoatrial
   - Third ventriculostomy (only in cases of obstructive hydrocephalus)[3,6]
9. *What are the main complications of these treatment measures?*
   - Complication rate of shunting in NPH is 30 to 40%. These include[3]:
     - Subdural hematoma (8–17%)
       - ○ Higher rate if the patient is older or if a low-pressure valve is used
       - ○ Two-thirds resolve spontaneously; one-third need evacuation and shunt tying
     - Shunt infection, obstruction, or disconnection (10–31%)
     - Intraparenchymal hemorrhage
     - Seizure (4%)
10. *Name some prognostic factors for the treatment of this condition.*
    - The most likely symptom to improve is incontinence, then ataxia, and lastly dementia.
    - There is a better response rate if the gait impairment is the primary symptom.[4,5]
    - Long-term response rate is as high as 75%.[4]
    - The response is better if the symptoms are present for a shorter time.[4]
    - Some patients (e.g., with Alzheimer's disease) will improve for a brief period and then worsen again.

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# Section VI Spine

# Case 99 Occipital Condyle Fractures

*Kelsey A. Walsh and Jason Tullis*

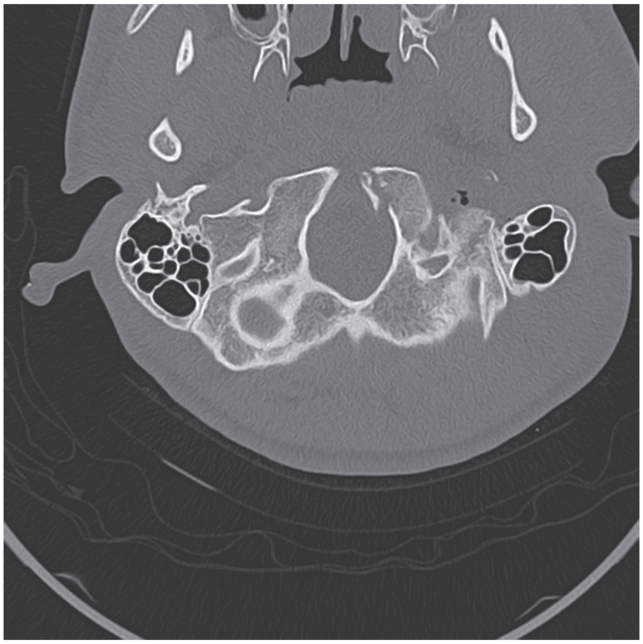


**Fig. 99.1** CT cervical spine, axial.

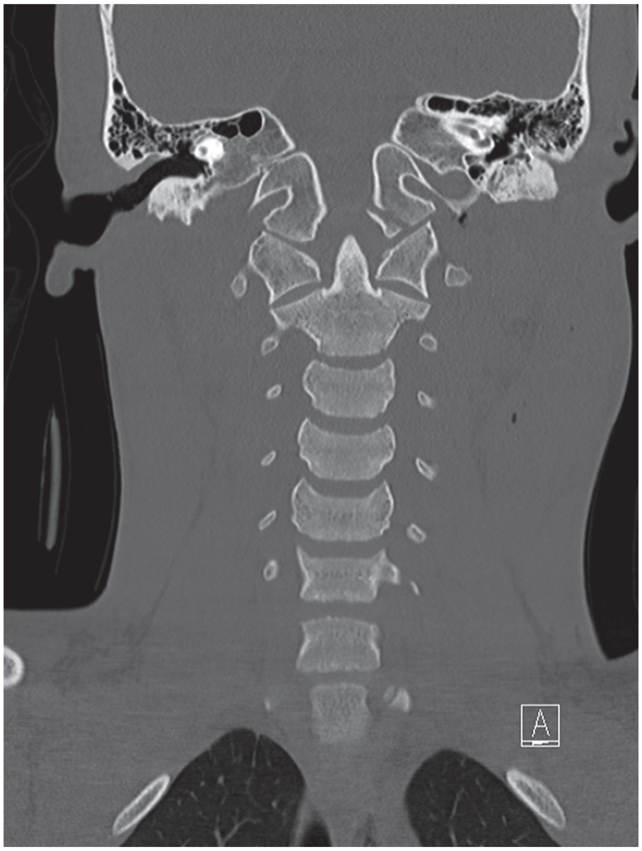


**Fig. 99.2** CT cervical spine, coronal.

## Clinical Presentation

- 25-year-old male presents to the emergency room following a motorcycle accident in which he was ejected from the motorcycle. The patient was wearing a helmet, but was intoxicated at the time.
- On exam, he is awake, alert, intoxicated, but oriented to person and place. He is complaining of headache and neck pain. His cranial nerves are intact and he has no gross motor or sensory deficits.
- Imaging is obtained in the emergency room (ER) and he is found to have facial fractures, traumatic subarachnoid hemorrhage, and multiple rib fractures, as well as the findings seen in ▶**Fig. 99.1** and ▶**Fig. 99.2.**

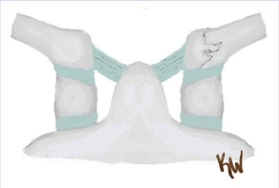
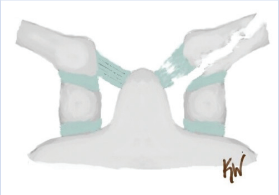
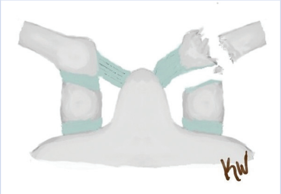
## Questions

1. Describe the CT scan.
2. What is the most commonly used classification system for occipital condyle fracture (OCF)?
3. Which type of fracture does this patient have?
4. Which is the most common type of OCF?
5. What is the typical clinical presentation for OCF?
6. How would you treat this patient?
7. How would your treatment change if this patient had bilateral OCF?
8. When should surgical intervention for OCF be considered?
9. The patient presents at your clinic 4 weeks later and has developed a cranial nerve deficit. Which cranial nerve is most likely injured?
10. In this patient with a left-sided OCF and an associated hypoglossal injury, in which direction would his tongue deviate on exam?

## Answers

1. *Describe the CT scan.*
    - Axial and coronal CT cervical spine imaging demonstrates an acute fracture of the left occipital condyle which appears to be comminuted with a fragment displaced medially (OCF).
2. *What is the most commonly used classification system for occipital condyle fracture (OCF)?*
    - Anderson and Montesano classification, see ▸**Table 99.1**
3. *Which type of fracture does this patient have?*
    - This patient has a left-sided type III OCF.
4. *Which is the most common type of OCF?*
    - Type III OCF comprise approximately 50% of all OCFs.[1]
5. *What is the typical clinical presentation for OCF?*
    - OCFs typically result from high-impact trauma in patients with multiple concomitant injuries, including traumatic brain injuries.
    - Most patients present with neck pain, and up to 40% may present with neurological deficits.[1]
        - Lower cranial nerve deficits are most common,[2] with some reported cases of Collet–Sicard syndrome,[3,4] nystagmus, or lateral rectus palsy.[5]
        - Patients may present with mono-, para-, or even quadriparesis.
    - Patients may develop delayed deficits, especially if untreated.
6. *How would you treat this patient?*
    - External immobilization with rigid cervical collar is the first line of treatment.
        - According to the AANS/CNS guidelines, all types of OCFs are recommended to be treated with C-collar immobilization initially, as it is almost always sufficient to prevent or treat a cranial nerve deficit and promote bony healing.[1]
    - An MRI should be obtained in order to assess ligamentous integrity.
        - Rotation or displacement of the occiput in relation to C1 requires evaluation for ligament injury and potential instability which may warrant further treatment (not present in this case).
7. *How would your treatment change if this patient had bilateral OCF?*
    - Immobilization with a halo vest device should be considered for bilateral OCFs.[1]
8. *When should surgical intervention for OCF be considered?*
    - Occipitocervical stabilization should be considered in any patients with bilateral OCF, OCF with clinical or radiographic evidence of occipitoatlantal or atlantoaxial instability,[1] or for decompression of a fracture fragment.[6]
9. *The patient presents at your clinic 4 weeks later and has developed a cranial nerve deficit. Which cranial nerve is most likely injured?*
    - CN XII, due to the close proximity of the hypoglossal canal to the occipital condyle
10. *In this patient with a left-sided OCF and an associated hypoglossal injury, in which direction would his tongue deviate on exam?*
    - A peripheral injury to the hypoglossal nerve causes tongue deviation toward the side of the injury. Thus, this patient's tongue would deviate to the left.[7]

**Table 99.1** Classification of occipital condyle fractures with illustrations showing a coronal view of the skull base, occipital condyle, C1 and C2 vertebral bodies in gray, with the alar ligament shown in blue

| | |
|---|---|
|  | **Type I**: Comminuted fracture due to axial loading injury. Minimal fracture displacement |
|  | **Type II**: Extension of a linear skull base fracture |
|  | **Type III**: Avulsion of condyle fragment, due to rotation or lateral bending injury |

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# Case 100 Jefferson Fractures

*Andrew Smith and Jason Tullis*

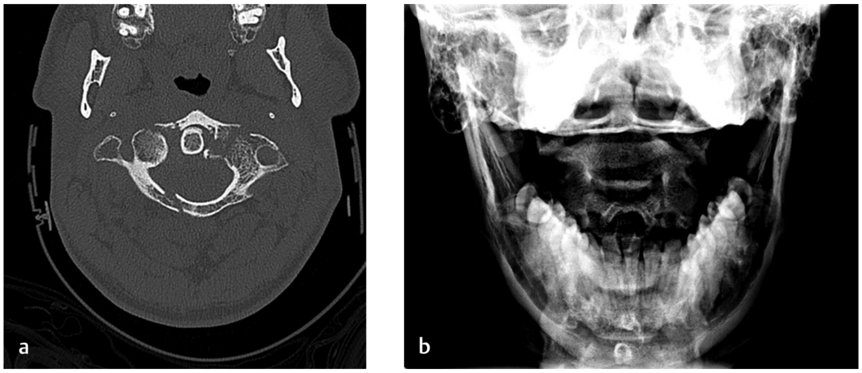

**Fig. 100.1** (**a**) Axial CT of the cervical spine at the level of the atlas. (**b**) Open mouth X-ray view of the odontoid.

## Clinical Presentation

- A 49-year-old male presents to the emergency department following a motor vehicle crash.
- He lost consciousness for approximately 5 minutes and is now complaining of significant posterior neck pain and stiffness. He denies numbness, tingling, weakness, or difficulty urinating.
- Patient has Glasgow Coma Score (GCS) of 15 with no appreciable motor or sensory deficits. Rectal sensation and voluntary contraction intact. Reflexes are 2+ throughout with no Hoffman, Babinski, or clonus.
- An open-mouth odontoid view plain film and a CT scan of the cervical spine are obtained and shown in ▶**Fig. 100.1** and ▶**Fig. 100.2**, respectively.

## Questions

1. Describe the images in ▶**Fig. 100.1**a, b.
2. Briefly describe the anatomy of the spinal column at the C1 level.
3. What is a Jefferson fracture? What are the different types of C1 fractures?
4. What symptoms are associated with C1 fractures?
5. What is the pathophysiology of a Jefferson fracture?
6. What other cervical spine fractures are commonly associated with C1 fractures?
7. What is the defining characteristic of an unstable isolated C1 fracture?
8. What are radiographic indicators for disruption of the transverse atlantal ligament (TAL)?
9. What are the management options for isolated C1 fractures?
10. What are the management options for combination atlas and axis fractures?

## Answers

1. *Describe the images in* ▶**Fig. 100.1**a, b.
   - The plain film open-mouth view shows an increased diameter between the medial edges of the lateral masses of C1 as well as > 7 mm of C1 lateral mass overhang on the lateral masses of C2.
   - The axial CT best demonstrates a fracture of both anterior and posterior arches.
   - There is also a comminution of the bilateral C1 lateral masses to involve the left atlanto-occipital joint and the bilateral atlantoaxial joints.
2. *Briefly describe the anatomy of the spinal column at the C1 level.*
   - The atlas is a ring of bone composed of two arches, anterior and posterior joined by the lateral masses.[1]

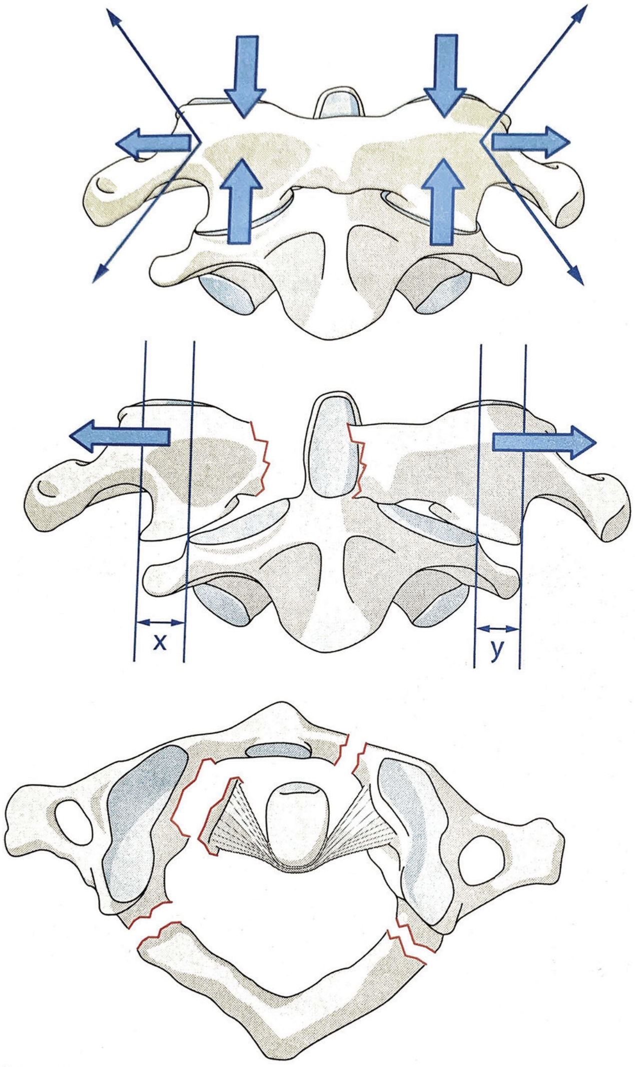

**Fig. 100.2** Illustration of mechanisms and force vectors applied to the spine leading to the occurrence of a Jefferson's Fracture. (Reproduced from AoSpine Manual: Principles and Techniques, Clinical Applications (2 Vol. Set) (v. 2) 1st ed. New York: Thieme; 2007)

## Answers (*continued*)

- Steel's rule of thirds states that the spinal cord and the odontoid process are each approximately 1 cm in diameter, approximately one-third the diameter of the ring of C1 of which the inner space is composed as follows: one-third spinal cord, one-third odontoid, and one-third space. The average diameter of the C1 canal is 3 cm giving 1 cm of "safe space."[2] A-P dislocation of C1 on C2 of more than 1 cm translates into possible spinal cord compression.[3]
- The TAL courses posteriorly to the dens attaching to the medial edges of the lateral masses of C1. It is the primary "defense" for anterior atlantoaxial subluxation.[1,2]
- The dens ligaments, the alar and apical, limit rotation. The alar ligaments become taught within the "safe space" preventing cord compression by the odontoid.[1] These are not strong enough alone to prevent posterior subluxation of the dens in an acute traumatic injury.[2]
- The greater occipital nerve exits under the posterior arch of C1 and runs superiorly to supply the V2 dermatome of the occipital scalp.[1]
- The suboccipital nerve exits above the posterior arch of C1.[1]
- The vertebral artery exits the transverse foramen, coursing posteriorly and crossing the arch at the junction of the lateral masses.

3. *What is a Jefferson fracture? What are the different types of C1 fractures?*
   - Acute atlas fractures comprise 2 to 13% of acute cervical spine fractures and 1 to 2% of all spinal column fractures.[4]
   - The classic Jefferson fracture as reported by Dr. Jefferson in 1920 is a four-point fracture involving the anterior and posterior rings.[5]
   - One of the commonly used classification systems of atlas fractures is the Landells classification.
     - Type 1: Isolated anterior or posterior arch fractures

## Answers (*continued*)

        - Most prevalent overall
        - Highest incidence of neurological deficit
        - Often associated with other cervical fractures
    - Type II: Anterior and posterior arch fractures
        - Often an isolated fracture
        - Most frequently unstable subtype
    - Type III: Lateral mass fracture
        - Least prevalent
- No established overall prognostic difference between the Landell fracture classifications
- There is a good overall prognosis for Jefferson fractures.[6]

4. *What symptoms are associated with C1 fractures?*
    - Neck pain[5]
    - Muscle spasm[5,7]
    - Occipital neuralgia and/or anesthesia to pin prick secondary to compression of the greater occipital nerve[2,5]
    - In severe cases, respiratory depression from compression of the medulla and potentially other long tract neurological signs[2]
5. *What is the pathophysiology of a Jefferson fracture?*
    - A classic Jefferson fracture typically occurs due to downward compression of the cranium such as from a heavy object falling on the patient's head. The force is transmitted from the cranium to the atlas articular facets through the occipital condyles. The remaining spinal column serves as natural upward resistance summing the force onto the C1 level.[5]
    - While initial logic dictates that the most common C1 injuries would occur to the articular facets and lateral masses, it is well known that the anterior and posterior rings are more commonly fractured. The triangular shape of the C1 lateral masses project the force from the occipital condyles down and out and the resistance force from the remaining spinal column up and out. This leaves the vector sum in a lateral direction accounting for the outward displacement that occurs in Jefferson fractures.[5] The weakest portion of the C1 vertebra is the junction of the anterior and posterior rings with the lateral masses.[1] These fracture first with disruption of the TAL in severe injuries and a resulting lateral displacement of the C1 lateral masses as viewed on open-mouth X-rays justifying the logic behind the rule of Spence. This outward displacement combined with the already increased canal diameter at the C1 level account for the rare incidence of spinal cord injury in Jefferson fractures (see ▶**Fig. 100.2**).
    - The mechanism of a C1 fracture of an isolated ring is thought to be due to hyperextension.[1]
    - The mechanism of a C1 lateral mass fracture is thought to be due to extreme force resulting in vertical compression with the head out of midline position.[1]
6. *What other cervical spine fractures are commonly associated with C1 fractures?*
    - Hangman's fracture
    - C2 body fractures
    - Odontoid fractures. C1-type II odontoid fracture is the most common C1–2 combination fracture. Type III odontoid fractures also occur.[5,8,9]
7. *What is the defining characteristic of an unstable isolated C1 fracture?*
    - Disruption of the TAL[10]
8. *What are radiographic indicators for disruption of the transverse atlantal ligament (TAL)?*
    - Rule of Spence: Sum of displacement of the lateral masses of C1 on C2 > 6.9 mm on open-mouth plain film.[11]
    - Increased atlantodental interval > 5 mm on plain radiograph.[3]
    - Abnormal signal in the TAL on MRI. In a study of 39 patients, Dickman et al reported an increased sensitivity for detection of disruption of the ligament over the rule of Spence (61% of disruptions would have been otherwise missed).[12]
    - Dickman et al also proposed a classification of TAL disruption:
        - Type I: Involve midportion of TAL or insertion of tubercle. Incapable of healing without external immobilization
        - Type II: Involve fractures and avulsion of tubercle for insertion of TAL. Capable of healing with external immobilization[12]
9. *What are the management options for isolated C1 fractures?*
    - No class I or class II medical evidence exists regarding management of these fractures.[10]
    - In cases where the TAL is intact (nondisplaced):
        - External immobilization devices such as rigid collars, suboccipital mandibular immobilizer braces, and halo ring vest orthoses for 8 to 12 weeks with follow-up plain films
        - Successful C1–C2 union was seen in 96% of nondisplaced type I and III fractures with no clear evidence to suggest one form of brace over another.[10]
    - In cases where the TAL is disrupted:
        - Initial reduction with traction followed by halo immobilization for combined total of 12 weeks.
        - Of 44 patients with isolated unstable C1 fractures treated with traction and rigid immobilization, 5 (16%) failed to achieve union, and 1 required surgical fusion.[6,7,13–15]
        - While osseous union with associated stability is achievable with conservative management, incomplete reduction resulting in long-term neck pain and stiffness can occur.[7,13]
        - The subsets by Levine et al showed union in all conservatively managed patients but the patients treated with traction for 1 week prior to

## Answers (*continued*)

immobilization lost reduction while patients treated with traction for 6 weeks maintained reduction.[7]

- Fowler et al also placed patients in traction for 6 weeks with a resulting average reduction of 50% of the initial displacement in 13 patients and complete reduction in only 3 (23%).[13]
- Incomplete initial reduction should favor internal reduction and fusion. Evidence of instability on radiographs 12 weeks after injury favor C1–C2 or occiput to C3 fusion.[10,15]
- One factor that could also favor early surgical intervention is a reportedly increased morbidity and mortality of halo stabilization in the elderly.[10]

10. *What are the management options for combination atlas and axis fractures?*
    - Combination atlas and axis fractures (level III):
      - Surgical stabilization and fusion is recommended when there are indications of spinal instability C1-type II odontoid combination fractures with atlantodental ratio of greater than or equal to 5 mm, C1-Hangman combination fractures with C2–C3 angulation of greater than or equal to 11 mm
      - Otherwise, cervical immobilization is recommended with follow-up plain films to assess union.[9]

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# Case 101 Hangman's Fracture

*Daniel S. Ikeda, Ahmed Mohyeldin, Evan S. Marlin, Hasel W. Slone, and H. Francis Farhadi*

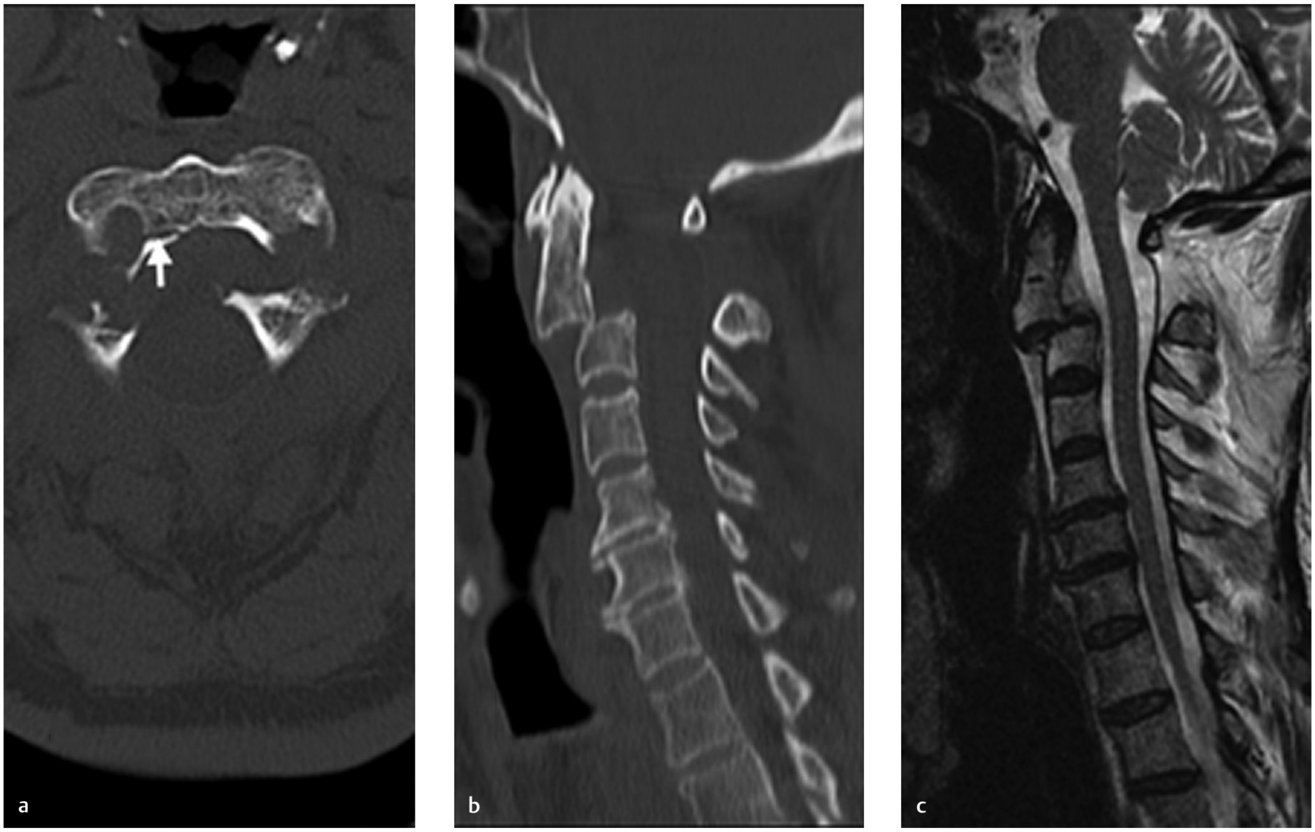

**Fig. 101.1** Axial (**a**) and sagittal (**b**) CT images demonstrate fractures through the bilateral pars interarticularis of C2 with associated anterior subluxation of C2 on C3. Note the fracture line extends through the right transverse foramen (*white arrow*). A sagittal T2-weighted MRI demonstrates disruption of the C2–C3 disc space with disruption of the posterior longitudinal ligament (**c**). Note the widening of the spinal canal.

## ■ Clinical Presentation

- A 66-year-old gentleman presents after a 200-lb barn door fell and struck his head and neck.
- He reports the door hit him in the forehead causing him to bend his neck "backward." The patient denies loss of consciousness but has had continued neck pain since the incident. He has no past medical history.
- On examination, his Glasgow Coma Score (GCS) is 15. There is a small laceration over his forehead, but no other injuries on primary or secondary trauma surveys.
- He has a normal neurological examination. A CT scan of the head is normal and a CT of the cervical spine is shown (▶**Fig. 101.1**a, b).

## Questions

1. What is the diagnosis?
2. How frequently do patients with these fractures suffer spinal cord injuries (SCI)?
3. What is your initial management?
4. What classification systems exist to help guide management in these injuries?
5. What are your options for treatment?
6. What are the strategies, contraindications, and potential pitfalls for patients treated with traction and rigid immobilization?
7. What are potential complications of surgical treatment?
8. How should we treat this patient?

## Answers

1. *What is the diagnosis?*
   - The CT scan demonstrates the typical appearance of a hangman's fracture or traumatic spondylolisthesis of the axis. The injury is a fracture through the bilateral pars interarticularis (isthmus) of the pedicle of C2 and is often accompanied by subluxation of C2 on C3.
   - It was first described in 1913 after autopsy examination of individuals executed by judicial hanging, although the term "hangman's fracture" was not coined until 1965 by Schneider et al.[1,2] The mechanism for fracture in judicial hangings is hyperextension and distraction, while most contemporary injuries are the result of hyperextension and axial loading.
     - Most injuries are incurred during motor vehicle collisions.
2. *How frequently do patients with these fractures suffer spinal cord injuries (SCI)?*
   - SCI is rare in these patients.
     - Most large series report an SCI incidence rate of 8% or less[3–5]
       - Patterns of injury include complete and incomplete tetraparesis, central cord syndrome, and Brown–Sequard syndrome.
       - As most contemporary injuries occur from hyperextension and axial loading, the spinal canal widens with the injury, thus protecting most patients from SCI.[6]
   - There is a high incidence of concomitant closed head injuries (up to 33%) and facial injuries (80%).[5,7]
     - In Effendi et al.'s series of 131 patients, the investigators found an associated all-cause mortality of 6.8%.
3. *What is your initial management?*
   - The patient should be admitted to the intensive care unit after thorough trauma evaluation.
   - Strict bed rest with spinal injury precautions is ordered and cervical orthosis should be applied.
   - An MRI of the cervical spine is ordered to evaluate the discoligamentous complex (DLC) and posterior ligamentous injuries (▶ **Fig. 101.1**c).
   - A CT angiogram (CTA) of the neck is ordered to evaluate the patient for any potential vertebral artery (VA) or other vascular injury
     - VA dissection is a potential cause of cerebral ischemic injury with possible neurologic morbidity and should be ruled out prior to any definitive treatment of the fracture.[3,8]
4. *What classification systems exist to help guide management in these injuries?*
   - Several classification schemes exist including the Effendi, modified Effendi, and Francis grading systems[3–5]
     - All classification schemes stress the importance of angulation of the body of C2, subluxation of C2 on C3, and integrity of the C2–C3 DLC.
     - It should be noted that the morphology of fractures and measurements for angulation were initially described based on plain radiographs.
   - The Effendi classification system, later modified by Levine and Edwards in 1985, is the most commonly used classification scheme (**Table 101.1**)[3,4] (▶ **Fig. 101.2**).
     - Type I injuries are felt to be **stable** as the fracture has spared the DLC and there is minimal subluxation and no angulation.
       - Nonsurgical management is appropriate and often employed
     - Type II and IIa injuries are generally felt to be **unstable** as the DLC of C2–C3 is disrupted.
       - There is significant variability in treatment depending on the degree of subluxation and/or angulation of C2 on C3.
     - Type III injuries are felt to be **unstable** as the fracture has led to disruption of the C2–C3 facet capsules in addition to the bilateral pars of C2.
       - Surgical management is appropriate.
5. *What are your options for treatment?*
   - Cervical orthosis is generally utilized for stable fractures (type I injuries).
     - Rigid immobilization with a halo vest can be used when there is concern for patient compliance or for type II injuries that are able to be reduced.

**Table 101.1** Levine and Edwards or modified Effendi classification system of hangman's fractures

| Type | Description | Fracture morphology | Comments |
|---|---|---|---|
| I | Vertical fractures through pars just posterior to C2 body | • ≤ 3 mm subluxation of C2 on C3<br>• No angulation | • Stable<br>• Neurological deficit rare |
| II | Type I + disruption of the C2–C3 DL complex | • > 3mm of subluxation<br>• ± Angulation | • Usually reduces with traction<br>• Can often be treated with rigid immobilization if closed reduction achieved |
| IIa | Oblique fracture through pars with DL disruption but competency of the anterior longitudinal ligament is often maintained | • Little subluxation (often ≤ 3 mm subluxation)<br>• Significant angulation (often > 15°) | • Unstable<br>• Traction should not be attempted if there is widening of the disc space |
| III | Type II + bilateral C2–C3 facet disruption | • Facets of C2–C3 subluxed or locked<br>• ± Subluxation of C2 on C3<br>• ± Angulation<br>• Mechanism may be from flexion and compression | • Unstable<br>• Neurological deficit more common<br>• Traction should not be attempted<br>• Requires posterior surgical fixation |

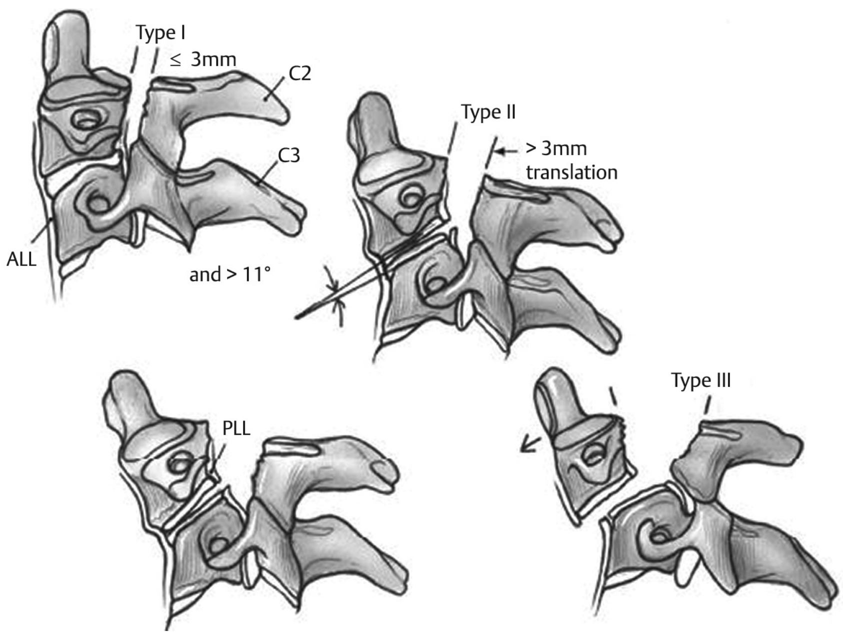

**Fig. 101.2** Classification of hangman's fractures: type I: ≤ 3 mm translation/no angulation; type II: > 3 mm of translation/> 11 degrees angulation; type III: severe angulation, translation with possible C2–C3 facet disruption. (Reproduced with permission from Neurosurgery Tricks of the Trade: Spine and Peripheral Nerve, Nader R., et al., Thieme New York; 2013).

## Answers (*continued*)

- Closed reduction with traction and rigid immobilization with a halo vest may be employed for displaced injuries without circumferential DL disruption and/or widening of the disc space.
- Surgical treatment is often employed for modified Effendi type IIa and III injuries (type II injuries are usually managed surgically if closed reduction fails or is aborted).
  - A C2–C3 anterior cervical discectomy and fusion (ACDF) is a viable option for type II and IIa injuries.
  - Various posterior cervical fixation techniques have also been reported with success.
    - C1–C2 wiring or lateral mass fixation with fusion (if there is significant DL disruption at C2–3, the fixation will need to extend to the C3 segment).
    - Direct osteosynthesis with screw placement across the fracture line for reduction and fixation (use in isolation without additional fixation constructs is contraindicated if DL disruption exists at C2–C3)

## Answers (*continued*)

- ○ Occipitocervical fixation if associated atlanto-occipital unstable injury

6. *What are the strategies, contraindications, and potential pitfalls for patients treated with traction and rigid immobilization?*
   - For decades, closed reduction with traction followed by rigid halo immobilization was the standard of care for many patients with hangman's fractures.
     - Some patients required 4 to 6 weeks of traction.[3,4,7]
       - ○ Given the advancements in surgical fixation techniques, risks of prolonged traction, and current views regarding prolonged hospitalization, patients are no longer treated in this manner.[9]
     - Advancements in MRI have revealed that fractures with severe DL disruption that should not be treated with traction.
   - Traction still has a role in contemporary treatment of some fractures, under the right clinical circumstances.
     - Traction can be done if a patient's neurological exam can be serially monitored. It should be limited to a duration of less than 7 days and should be aborted or discontinued immediately if there is a change in the neurological exam or radiographic evidence of distraction of the C2 body.
     - Traction should be started at 10 lb with the patient in slight extension.
       - ○ Weight can then be increased in 5-lb increments up to a maximum of 30 lb. (20 lb is usually sufficient).
     - Closed reduction with traction is contraindicated in patients with severe angulation (type IIa injuries), severe DL disruption, and type III injuries.
7. *What are potential complications of surgical treatment?*
   - Anterior surgery for hangman's fractures require at least a C2–C3 ACDF and sometimes a C3 corpectomy if there is disruption of the C3 body.
     - A retropharyngeal approach is required and neurovascular structures, including the hypoglossal and superior laryngeal nerves, are at risk and must be protected.
     - In the largest series of anterior cervical surgery for hangman's fractures, investigators found rates of complications comparable to other indications for ACDF, including postoperative hematoma (5.3%), transient neurological deficit (7.9%), hoarseness (5.3%), and dysphagia (13.2%).[10]
     - Fusion rates have been reported to be as high as 95%, solidifying this approach as a viable option in appropriately selected patients.
   - A variety of posterior cervical fixation techniques may be utilized for these fractures.
     - A rotating spinal frame for prone positioning (so as to not lose reduction or cause a neurological injury) is recommended.
     - Preoperative CTA is invaluable in identifying the dominant VA and possible variant VA anatomy.
     - If screw purchase across the fractured isthmus is desired, intraoperative imaging with fluoroscopy or navigation is recommended as these fractures can be highly mobile and the fracture lines are variable.
     - Delayed instability has been reported to be higher with posterior approaches and fusion across the C2–C3 joint is recommended.[11] In cases requiring occipital cervical fusion, it is important that the patient be told that approximately 50% of head rotation will be lost with fixation across C1–C2.
8. *How should we treat this patient?*
   - The patient has a modified Effendi type II injury.
   - Closed reduction with gentle traction was attempted, but this failed after 48 hours and the patient was unable to tolerate any further traction.
   - A C2–C3 ACDF was performed with bicortical screw fixation, followed by rigid cervical orthosis application for 3 months (see ▸**Fig. 101.3**).
     - The patient had temporary dysphagia postoperatively and needed nasogastric (NG) tube feeds for 1 week.
     - Delayed flexion and extension cervical X-rays demonstrated solid fusion without any instability.

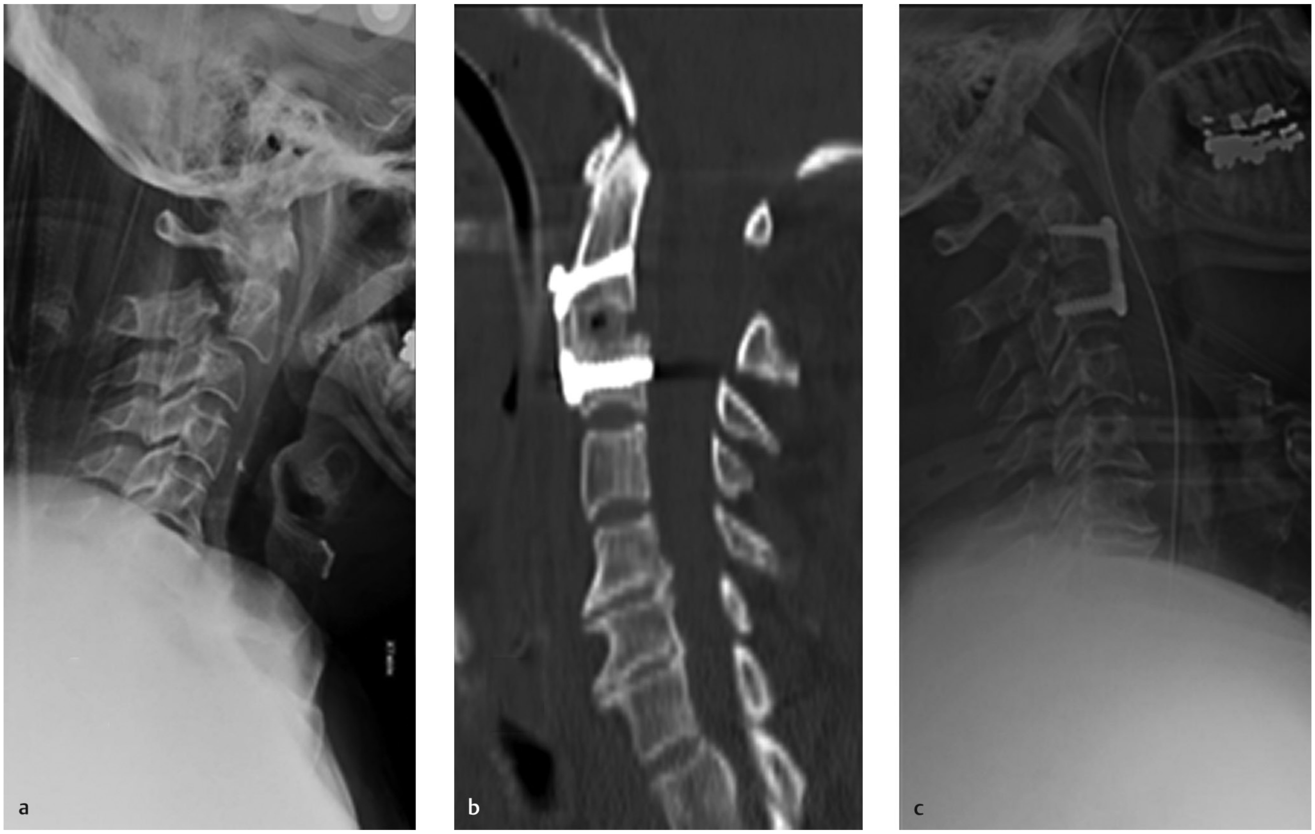

**Fig. 101.3** A lateral cervical X-ray demonstrates failure to reduce the fracture with traction (**a**). A postoperative CT image demonstrates bicortical screw purchase for fixation and restoration of alignment (**b**). A postoperative lateral cervical X-ray demonstrates realignment of the pars interarticularis with improved alignment (**c**).

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# Case 102 Atlantoaxial Instability

*Jeffrey P. Mullin, Alvin Chan, Eric P. Roger, and Edward C. Benzel*

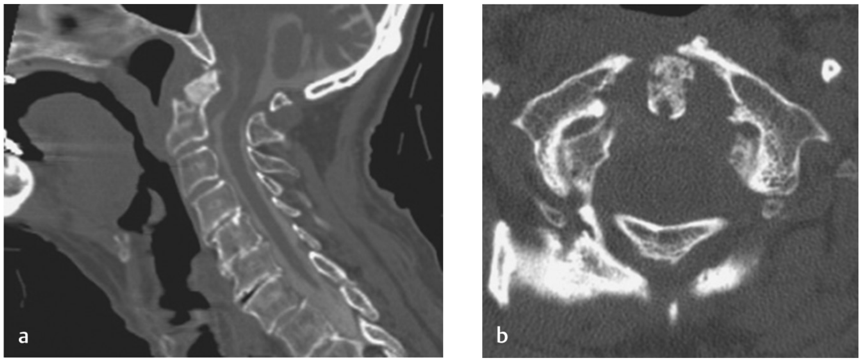

**Fig. 102.1** (**a**) Sagittal CT reconstruction with myelographic subarachnoid contrast injection. (**b**) Axial CT nonmyelogram cut at the level of C1 through the dens.

## Clinical Presentation

- An 83-year-old man presents with neck pain.
- He sustained a fall 6 months prior. There was no documentation of fracture at the time. He has been experiencing neck pain since the fall.
- He denies any bladder or bowel dysfunction, gait or balance disorders.
- He is admitted for generalized weakness and shortness of breath.
- Upon further work-up, he is found to be severely hyperkalemic with mental changes.
- His past medical history is remarkable for placement of a cardiac pacemaker.

## Questions

1. What would be the sequence of radiologic investigations recommended for this patient?
2. Interpret the images in ▶**Fig. 102.1**.
3. Is this an acute or chronic process? Why?
4. How commonly are the pathologies at C1 and C2 associated?
5. What are your therapeutic options at this point? How likely are they to be successful in achieving stability?
6. Is spinal cord decompression indicated?
7. What techniques are available for C1–C2 fixation? What factors would influence the decision-making?
8. Would an odontoid screw be an appropriate surgical option in this patient?
9. List the disadvantages of posterior C1–C2 fusion.
10. What are the pros and cons of transecting the C2 nerve root when placing a C1 lateral mass screw?
11. If an arcuate foramen is shown on preoperative imaging, what is a potential complication of placing a C1 lateral mass screw and how could it be avoided?

## Answers

1. *What would be the sequence of radiologic investigations recommended for this patient?*
   - The patient should be investigated with the following:
     - Chest radiograph, as the patient is short of breath
     - Plain anteroposterior (AP) and lateral cervical radiographs
     - Due to his pacemaker, MRI is not possible.
     - CT myelogram is the best “second choice” instead of an MRI to assess the status of spinal cord and soft tissues (anterior soft tissue swelling and pannus). You may skip the CT myelogram if the patient is not myelopathic.
     - CT scan is best to assess bony integrity and bony anatomy. It is highly indicated preoperatively for atlantoaxial stabilization.
     - Flexion/extension radiographs should never be performed when previous imaging shows evidence of instability. These should only be used to confirm previously negative/normal imaging.[1]

## Answers (*continued*)

2. *Interpret the images in* ▶**Fig. 102.1**.
   - Type II odontoid fracture with minimal translation, but ~60 degrees of posterior angulation
   - Soft tissue pannus behind the odontoid, partially indenting the cerebrospinal fluid (CSF) space, but not in direct contact with the cord (at least in this static CT image in supine position)
   - Axial image demonstrates anterior atlantal ring fracture consistent with a Jefferson fracture (posterior ring fracture not illustrated). This finding is suggested on ▶**Fig. 102.1**a by the absence of the anterior ring on this midline cut.
   - Various levels of degenerative changes are noted through the rest of the cervical spine; there is no evidence of cord compression.
3. *Is this an acute or chronic process? Why?*
   - These fractures are likely to be chronic because of the following:
     - The fracture line at the base of the dens appears sclerotic.
     - The fracture of the anterior atlantal arch is completely corticated.
     - Soft tissue pannus formation behind the odontoid is suggestive of chronic instability.
     - The patient presents with a fall 6 months prior, with neck pain since. There is no clinical evidence of an acute fall (i.e., don't forget the clues given on history!).
4. *How commonly are the pathologies at C1 and C2 associated?*
   - Axis fractures are relatively commonly associated with C1 fractures. Odontoid fractures (type II or III) are associated with C1 fractures in up to 53% of cases, and up to 26% with hangman's fractures. They are reported to have a higher morbidity and mortality rate. Treatment is primarily based on the characteristics of the C2 fracture.[2]
5. *What are your therapeutic options at this point? How likely are they to be successful in achieving stability?*
   - Conservative management:
     - Soft collar
     - Semirigid collar (Philadelphia, Aspen, Miami J, etc.)
     - Rigid cervicothoracic orthosis
       - Minerva brace
       - Halo
   - Surgical stabilization[3]
     - Occipitocervical fusion
     - Atlantoaxial fusion
     - Odontoid screw is *not* an option in chronic fractures.
   - As these fractures are chronic, they are highly unlikely to heal with any type of external immobilization. Only open fixation and bone grafting can achieve stability. Conservative management should be considered only for patients who are medically unfit for surgery or, obviously, for patients refusing surgery.
   - Halo bracing is poorly tolerated by geriatric patients. Furthermore, although solid fixation of the head and chest may be achieved, paradoxical "snaking" motion of the cervical spine may occur.
   - Open fixation with bone grafting has a high fusion rate (> 90%), except in smokers or in patients on steroids.
6. *Is spinal cord decompression indicated?*
   - No. As illustrated by the CT myelogram, although there is soft tissue pannus formation with indentation onto the CSF space, there is no active cord contact or compression.
   - Furthermore, the patient does not present with any signs of myelopathy.
7. *What techniques are available for C1–C2 fixation? What factors would influence the decision-making?*
   - Available techniques include the following[3–5]:
     - Spinous process wiring (requires a structurally intact arch of C1):
       - Gallie fusion
       - Brookes fusion
       - Modified Sonntag fusion
     - C1–C2 fixation:
       - C1 and C2 pars screws
       - C1 pars and C2 pedicle screws
       - C1 pars and C2 translaminar/interlaminar screws
       - C1–C2 transarticular screws
       - C1–C2 Halifax clamps
     - Odontoid screw placement
     - Occiput to C2 fixation
   - C1–C2 spinous process wiring (Brooks, Gallie, Sonntag) is a good adjunct to transarticular screws or to a segmented fusion but is not biomechanically a very strong construct when used as standalone.[6]
   - Spinous process wiring may be performed in a variety of ways but requires the integrity of the lamina and spinous process of C2, as well as integrity of the posterior arch of C1. In this case, although no decompression is required, there is loss of integrity of the arch of C1 due to the Jefferson fracture.
   - C1–C2 transarticular screw placement and C1–C2 segmental fusion (C1 lateral mass screws, C2 pars interarticularis screws/C2 pedicle screw fixation) in conjunction with spinous process wiring results in biomechanically strong constructs with higher rates of successful fusion when compared with spinous process wiring alone.[7]
   - C1–C2 posterior fixation techniques do not rely on the integrity of the posterior elements. Nonetheless, they are not without risk. Specifically, the trajectory of the vertebral artery and bony anatomy of the C2 pars must be carefully studied on preoperative imaging. High-riding vertebral arteries or a thin C2 pars may preclude the placement of a transarticular screw (▶**Fig. 102.2** for ideal screw trajectory). If the arches of C1 or C2 are compromised (making spinous process wiring procedure not feasible), an

## Answers (*continued*)

onlay fusion laterally at the level of the articulating processes may alternatively be used in conjunction with C1–C2 transarticular screw placement.

- C2 fixation using translaminar/interlaminar screws significantly reduces the risks to the vertebral artery, although epidural cortical breach is possible.
- Preservation of C1–C2 rotation can also be achieved with a halo; however, this comes with associated morbidity in the elderly population.[8]

8. *Would an odontoid screw be an appropriate surgical option in this patient?*
    - An odontoid screw would not be a good option in this case. Odontoid screws are indicated for acute type II odontoid fractures only. The screw is intended to "pull" the free dens against the C2 body, encouraging healing by compression along the fracture line.
9. *List the disadvantages of posterior C1–C2 fusion.*
    - Disadvantages of C1–C2 posterior fixation include the loss of 50% of head rotation.
    - Furthermore, there is the potential for vertebral artery injury as well as significant venous plexus bleeding during lateral dissection of the C1–C2 joint.
    - The vertebral artery may be located in the path of a transarticular screw on preoperative images on at least one side in 18 to 23% of patients.[9]
    - Rate of intraoperative vertebral artery injury with this approach is estimated to be 1.7 to 4.1%.[10]
    - When C1–C2 spinous process wiring is utilized as a stand-alone technique, there is a high rate of pseudoarthrosis approaching 30% without halo fixation.[10]
    - Posterior C1–C2 instrumentation has been reported as a significant cause of postoperative occipital neuralgia.[11]
10. *What are the pros and cons of transecting the C2 nerve root when placing a C1 lateral mass screw?*
    - Transecting the C2 nerve root allows for: (1) better visualization of the C1 lateral mass, (2) better access to the C1–C2 articulation and C2 foramen for bone graft placement, and (3) decreased bleeding associated with the vascular plexus that surrounds the C2 nerve root.[12]
    - Intentionally sacrificing the C2 nerve root while placing a C1 lateral mass screw often has potential additional benefits like shorter OR times and comparable patient satisfaction scores in comparison to C2 nerve root preservation.[12]
    - A potential con for transecting the C2 nerve root is that occipital neuralgia may develop,[13] though there is evidence that transecting the C2 nerve root actually precludes occipital neuralgia.[14]
11. *If an arcuate foramen is shown on preoperative imaging, what is a potential complication of placing a C1 lateral mass screw and how could it be avoided?*
    - An arcuate foramen (i.e., ponticulus posticus) is a relatively common anatomical variant where a bony bridge covers the groove of C1 that holds the vertebral artery.
    - The arcuate foramen should be avoided as the starting point for a lateral mass screw to prevent lacerating the vertebral artery and thus potential bleeding.[15]
    - If vertebral artery injury is encountered, bleeding can be stopped via tamponade from placing the screw. If this is the first screw (and you still need to do the other side) you should *not* place the second screw for fear of causing bilateral vertebral artery damage potentially leading to devastating neurologic sequelae including death. Unilateral fixation is then accepted. Spinous process wiring may be considered.[10]
    - If such bleeding occurs while working on the second screw, then placement of the screw is again the only way to stop the bleeding.
    - Postoperative vertebral angiogram is indicated if damage to the vertebral artery is suspected. Vertebral artery stenting may be considered.

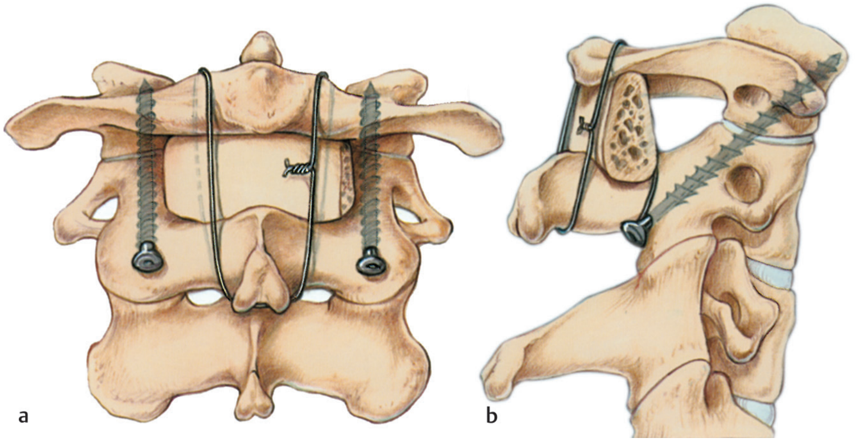

**Fig. 102.2** Artist's rendering of ideal trajectory of C1–C2 transarticular screws with (**a**) anteroposterior and (**b**) lateral illustrations. Posterior wiring with interspinous iliac graft is also depicted. Note that the posterior wiring of C1 to C2 may require a structurally intact arch of C1 and may not be feasible in some cases of Jefferson fracture (which is possible in this case). (Reproduced from Wolfla CE, Resnick DK. Neurosurgical Operative Atlas. Spine and Peripheral Nerves. New York: Thieme/American Association of Neurological Surgeons; 2006.)

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# Case 103 Type 2 Odontoid Fracture

*Christopher Evan Stewart, Joseph A. Shehadi, and Brian Seaman*

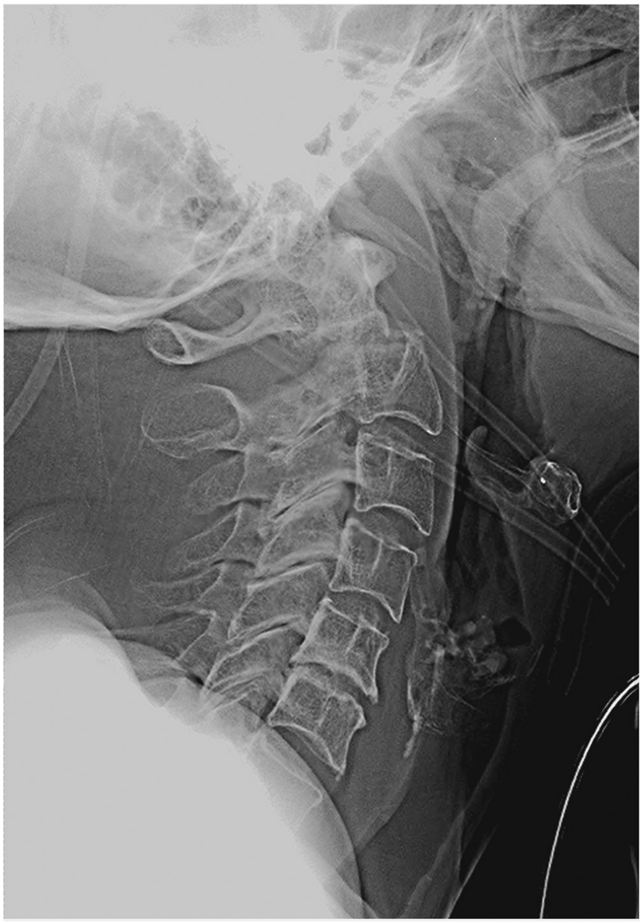

**Fig. 103.1** Lateral scout view of cervical spine CT scan.

## ■ Clinical Presentation

- A 77-year-old woman presents after a fall down five stairs. She complains of nonradiating neck pain.
- Her neurologic exam is unremarkable.
- You are asked to consult on this patient regarding a cervical spine fracture seen on a CT scan of the cervical spine (▶**Fig. 103.1**).

## ■ Questions

1. Interpret the CT scan of the cervical spine (▶**Fig. 103.1**).
2. Are there any additional studies you would order and why?
3. Odontoid fracture displacement and angulation are known to be important prognostic factors of fracture healing. Classification of these fractures significantly affects management decisions. Describe the classification system of Anderson and D'Alonzo for odontoid fractures including implications for spinal stability and indications for operation.

It was decided that surgical intervention is indicated for this acute type II odontoid fracture. MRI was performed and the transverse ligament was intact

## Questions (*continued*)

(▶**Fig. 103.2**). We elected against halo placement in this patient given the patient's age, associated comorbidities, and the higher risk for nonunion with a halo.

4. Name some contraindications to anterior odontoid screw placement.
5. Explain how you would reduce the fracture segment to achieve osseous contact prior to anterior odontoid screw fixation.
6. Highlight the key procedural steps for anterior odontoid screw placement.

The patient successfully underwent placement of a single odontoid lag screw under general anesthesia (▶**Fig. 103.3**). The use of one screw is thought to have similar biomechanical strength as two screws side by side.[1] Successful reduction of the fracture segment was obtained during patient positioning. The patient was placed in a hard collar for 3 months. She remained neurologically intact and without neck pain. Follow-up radiographs demonstrated good bony union of the fracture segment.

7. If in the immediate postoperative period following anterior odontoid screw placement your patient experiences increased odynophagia, coughing, and persistent fever, what complication must you have a high suspicion for?
8. Assume you treat a type II odontoid fracture with halo fixation. After 3 months of fixation, the patient develops recurrent neck pain. Flexion–extension radiographs reveal abnormal motion of the dens fragment, indicating a nonunion. How would you proceed?
9. How would you manage a patient with an odontoid screw breakage first noted on the 2-month postoperative radiograph?

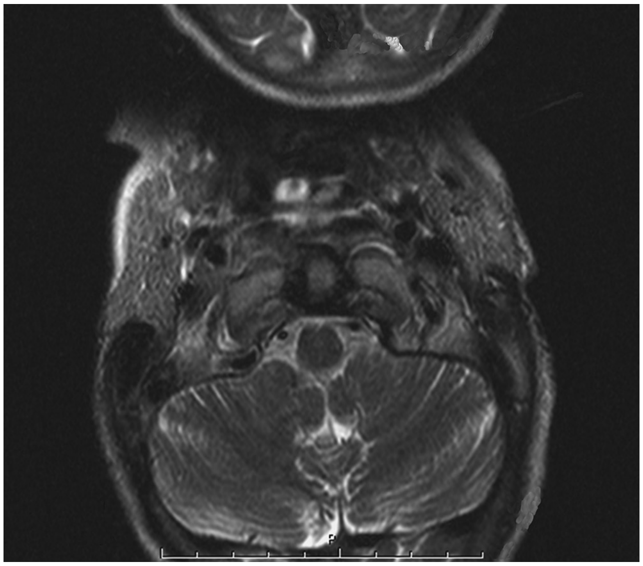

**Fig. 103.2** Axial MRI scan of cervical spine demonstrating transverse ligament.

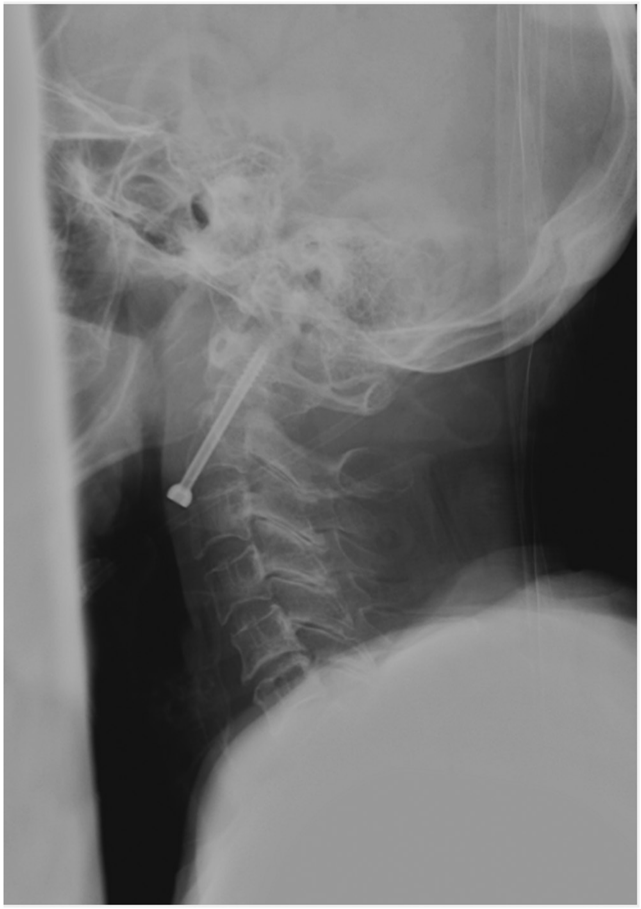

**Fig. 103.3** Postoperative plain lateral cervical spine radiograph demonstrating odontoid screw in place.

## Answers

1. *Interpret the CT scan of the cervical spine* (▶**Fig. 103.1**).
   - The lateral scout view of the CT scan of the cervical spine demonstrates a fracture of C2 at the base of the odontoid process consistent with an acute type II odontoid fracture.
   - There is slight posterior angulation of the dens.
   - The fracture has an oblique anterior–superior to inferior–posterior orientation.
   - The atlantodens interval is within normal limits.
2. *Are there any additional studies you would order and why?*
   - MRI is indicated to assess the status of the transverse and alar ligaments as well as the tectorial membrane. The integrity of the transverse ligament in addition to the atlantoaxial distances and relationships significantly affects your management and operative approach.[2]
3. *Odontoid fracture displacement and angulation are known to be important prognostic factors of fracture healing. Classification of these fractures significantly affects management decisions. Describe the classification system of Anderson and D'Alonzo for odontoid fractures including implications for spinal stability and indications for operation.*
   - Anderson and D'Alonzo's classification is described below (▶**Fig. 103.4**).[3]
     - Type I fractures occur through the tip of the dens above the transverse ligament. These are rare injuries and associated occipital–cervical dislocation should be excluded.[4]
     - Type II fractures occur through the base of the neck and are the most common fracture subtype.
     - Type IIA fractures are similar to type II, but there is comminution and large bone fragments at the fracture site.
     - Type III fractures occur through the body of C2 and may involve the articulating facet and marrow space. Type III fractures may be better characterized as horizontal rostral C2 body fracture and not odontoid process fractures.[5]
   - Type I and III fractures are often treated in a conservative fashion.[3] That said, one must consider "shallow" or "high" type III odontoid fractures as potentially unstable fractures.[4,6] Furthermore, anteriorly displaced type III fractures may also be unstable.
   - Attention should be given to the extent of articulating facet injury/distraction, which can be seen with these injuries.[7]
   - It is generally accepted that advanced age, fracture displacement > 6 mm, and angulation > 10 degrees negatively influences union rates.[4,8] Other factors include chronicity of the fracture,[9] delay in diagnosis, fracture comminution,[4] or inability to maintain fracture alignment with external immobilization.[10]
   - Type II odontoid fractures in patients 50 years and older should be considered for surgical stabilization and fusion.[10] This is based on a 21 times higher rate of nonunion in patients over the age of 50 years.[11]
4. *Name some contraindications to anterior odontoid screw placement.*
   - Contraindications for anterior odontoid screw fixation include the inability to reduce anatomically the fracture, nonunion, severe osteoporosis, transverse atlantal ligament rupture, or concomitant Jefferson type fracture with coronal plane separation of > 7 mm, or oblique fracture from anteroinferior to posterosuperior.[12]
   - Furthermore, this procedure is difficult in patients with short necks, barrel chests, those unable to tolerate cervical extension (spinal stenosis), or those with tracheostomies or significant open trauma to the anterior aspect of the neck.
5. *Explain how you would reduce the fracture segment to achieve osseous contact prior to anterior odontoid screw fixation.*
   - Typically, reduction of an odontoid fracture is obtained under fluoroscopy during operative positioning in the supine position with the Mayfield head holder.
   - Alternatively, Gardner–Wells tongs or manual cervical traction can be utilized to obtain reduction under fluoroscopy. Bivector traction with a flexor component is also an option. Close monitoring of neurological status should be obtained while performing any type of reduction; this may be done with serial neurological examinations while the patient is kept awake or by using intraoperative neuromonitoring such as motor and somatosensory evoked potentials.
6. *Highlight the key procedural steps for anterior odontoid screw placement.*
   Details of the procedure are described below[13] and are illustrated in ▶**Fig. 103.5**.[14]
   - The patient is positioned supine. The Mayfield head holder is utilized and the head is fixed in extension. Alternatively, intraoperative traction with Gardner–Wells tongs is utilized.
   - High-resolution biplanar fluoroscopic imaging is utilized to ensure reduction of the anterior displaced fracture segment and guide screw placement.
   - An anteromedian neck incision is placed in the region of C5–C6.
   - An avascular plane is dissected cephalad until the anteroinferior border of C2 is reached.[15]
   - A Hohmann retractor may be used to aid in exposure. Then, a high-speed drill is used to make a trough in the superior portion of C3 to allow for a proper angle when drilling a pilot hole.

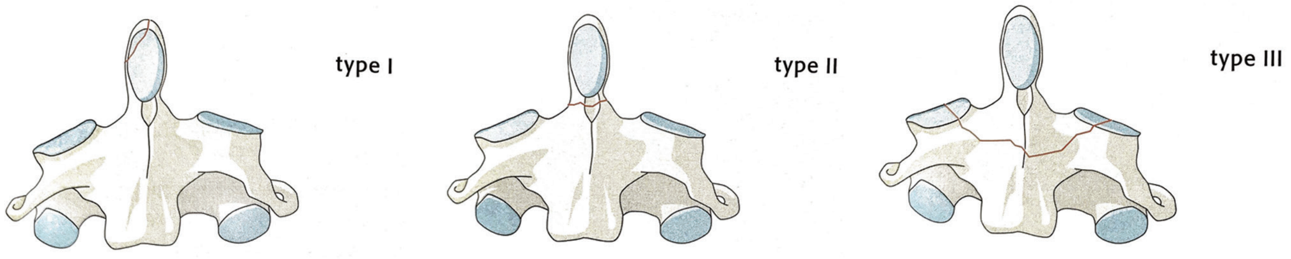

**Fig. 103.4** Classification of odontoid fractures. (Reproduced with permission from AoSpine Manual: Principles and Techniques, Clinical Applications (2 Vol. Set) (v. 2) 1st ed. New York: Thieme; 2007).

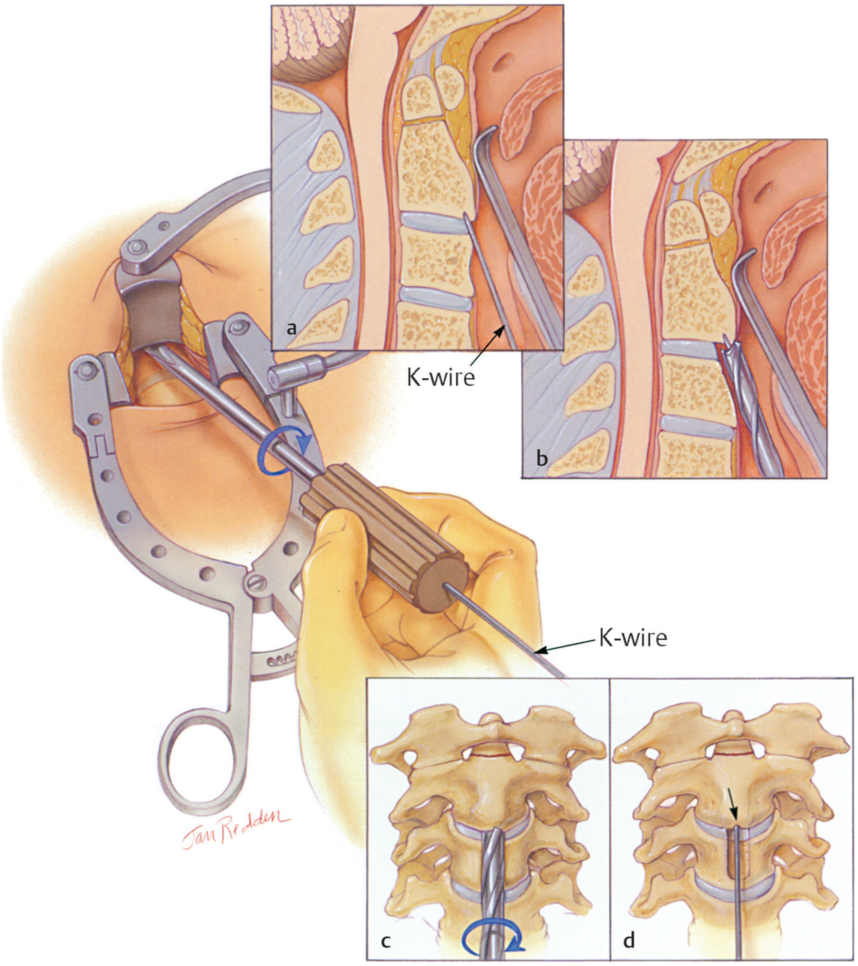

**Fig. 103.5** Artist's rendering of odontoid screw placement technique. (**a**) K-wire is placed entering at the anteroinferior edge of C2 and aimed toward the odontoid tip under fluoroscopy. (**b**) This is followed by drilling a hole core over the K-wire. (**c, d**) Part of the C2–C3 anulus is removed with the coring drill. (Reproduced with permission from Wolfla and Resnick 2006).[14]

## Answers (*continued*)

- Under fluoroscopic guidance, a K-wire is inserted and then a long 2.5-mm cannulated drill is inserted into the trough and angled posterior to reach the dorsal portion of the odontoid tip.
- On the anteroposterior projection midline trajectory is confirmed.
- The pilot hole is drilled, and the depth of the hole is measured.
- Then the appropriate-length 3.5-mm cannulated lag screw is placed through the pilot hole to obtain bicortical purchase.[16,17] Compression of the fracture segment should be seen on X-ray.

7. *If in the immediate postoperative period following anterior odontoid screw placement your patient experiences increased odynophagia, coughing, and persistent fever, what complication must you have a high suspicion for?*
   - Continued pain, fevers, odynophagia, incision swelling, drainage and coughing are common signs and symptoms of esophageal injury.[18]

## Answers (*continued*)

- Diagnosis is aided by contrasted swallow study, endoscopy, cervical radiographs, and oral methylene blue.[18]
- Appropriate and prompt investigation and treatment is essential for acceptable patient outcomes.

8. *Assume you treat a type II odontoid fracture with halo fixation. After 3 months of fixation, the patient develops recurrent neck pain. Flexion–extension radiographs reveal abnormal motion of the dens fragment, indicating a nonunion. How would you proceed?*
    - The development of high cervical neck pain or signs of myelopathy may signal nonunion.
    - An anterior odontoid screw is not recommended at this time secondary to fibrous scar, which is present at the fracture site.
    - In this situation, a posterior C1–C2 fusion would be indicated.[4,19,20]
9. *How would you manage a patient with an odontoid screw breakage first noted on the 2-month postoperative radiograph?*
    - Screw breakage may be a sign of nonunion. Initially, a CT scan of the cervical spine and flexion–extension X-rays should be performed. If a clear lucency is appreciated near the fracture site or if there is movement of the fracture segment, then a posterior cervical C1–C2 fusion should be considered.[19,20]

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# Case 104 Basilar Invagination

*Michel Lacroix*

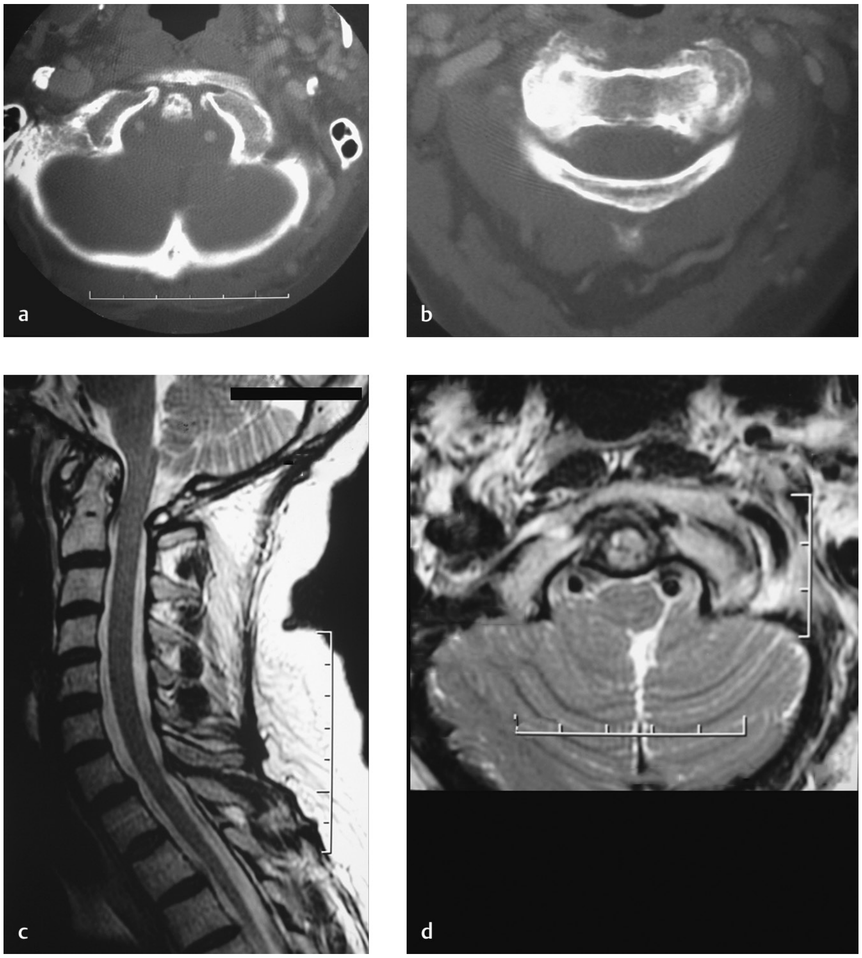

**Fig. 104.1** Cervical spine imaging with **(a)** axial CT at the level of the foramen magnum and **(b)** at the atlantoaxial junction. **(c)** T2-weighted sagittal MRI and **(d)** axial MRI at the level of the foramen magnum.

## Clinical Presentation

- A 55-year-old woman with rheumatoid arthritis presents with neck pain and progressive difficulty ambulating.
- She suffers from bilateral limb paresthesias. She is showing signs of myelopathy with hyperreflexia and bilateral Babinski's signs.
- CT scan and MRI of the cervical spine are obtained (▶**Fig. 104.1**).

## Questions

1. Are there any other symptoms or signs you would like to identify?
2. Interpret the CT and the MRI scans.
3. What is your initial management?
4. What studies do you order?
5. What is your course of action?

   After gradually putting her in 12 pounds of traction and mild sedation for 4 days, the CT scan shows visible reduction.
6. Describe the surgical options and select the one you would prefer.

   A posterior occipitocervical fusion, in situ distraction with transarticular C1–C2 screws and iliac crest autograft fusion was performed. A postoperative radiograph is shown in ▶**Fig. 104.2**. Solid fusion was observed and myelopathy improved.
7. After posterior fusion, what is the natural history of the basilar invagination?

## Answers

1. *Are there any other symptoms or signs you would like to identify?*
   - Hypoglossal, glossopharyngeal, vagus, and trigeminal nerve deficit and neurogenic bladder are signs of severe dysfunction at the craniovertebral junction.[1,2]
   - The accentuation of the symptoms in flexion and extension may suggest an atlanto-occipital segment hypermobility.
2. *Interpret the CT and the MRI scans.*
   - On the axial CT scan, the erosion and compression of the lateral atlantal masses, the atlantoaxial subluxation as well as the penetration of the dens beyond the level of the occipital condyle in the foramen magnum are visualized. The sagittal T2-weighted MR image of the cervical spine clearly shows some typical findings.[3,4]
     - The downward separation of the anterior arch of the atlas from the clivus provokes a descent (or telescopic scissoring) of the atlas arch onto the axis body and concomitant upward displacement of the dens.
     - The displacement of the posterior arch of the atlas rostrally and ventrally causes a decrease in the anteroposterior diameter of the spinal canal.
   - The inflammatory rheumatoid pannus of the odontoid process (synovial joints anteriorly with C1 and posteriorly with the transverse ligament) and the resulting compression of the medulla are seen in both the axial and sagittal MRI. There is no syringomyelia or Chiari malformation. The craniometric lines[3,5] in lateral view are shown in ▶**Fig. 104.3**.
3. *What is your initial management?*
   - Marks and Sharp[2] showed in 1981 that untreated myelopathic rheumatoid patients died within 6 months of presentation. Furthermore, most myelopathic rheumatoid patients (56%, $n = 18$) treated with conservative measures only (collar or traction alone) also died within 6 months.[2]
   - Treatment should be planned immediately.
4. *What studies do you order?*
   - Spinal and complete systemic work-up are in order:
     - Flexion–extension cervical spine X-rays. Any increase in the atlantodental interval confirms the instability, which is consistently associated with the basilar invagination.[1,3,4]
     - Consider dynamic MRI in flexion and extension to identify instability and sites of compression.
     - Preoperative blood work including nutritional status (lymphocyte count and liver function tests).
     - Discontinue nonsteroidal anti-inflammatory agents 1 week prior to surgery. Methotrexate and steroids may be used for pain management preoperatively.
     - Cardiac status, pulmonary function assessment tests, and anesthesia consult should be obtained.
5. *What is your course of action?*
   - A period of 4 to 5 days of halo traction with mild sedation and a maximum of 12 pounds of charge can reduce a basilar invagination of less than 15 mm.[4]
   - Traction is contraindicated in posterior occipitoatlantal dislocation or complex rotatory luxations.
6. *Describe the surgical options and select the one you would prefer.*
   - Treatment should be designed depending on the presence of an irreducible versus reducible deformity, stability and motion dynamics and site of encroachment.[4,6,7]
     - Irreducible deformity: The approach depends on the type of encroachment. If the encroachment is dorsal, a dorsal decompression is warranted. If the encroachment is ventrolateral, a ventral or anterior decompression is the treatment of choice. The anterior decompression can be ideally achieved through a transoral or transcervical route if the invagination is below the hard palate (level of oropharynx). If the invagination reaches above the hard palate, an endonasal or transoral route is preferred. Endoscopic transcervical[8] or transsphenoidal[7] approach instead of open approaches have also been described. The

## Answers (*continued*)

decompression is followed by a posterior fusion when the instability has been demonstrated, following the same approach described for the reducible deformity fixation below.

- Reducible deformity: Posterior occipitocervical fusion in most circumstances unless there is no demonstration of atlanto-occipital instability (clivus-occipital condyle and C1 arch relationship) and when a C1–C2 fusion only would be appropriate. Many posterior fusion techniques have been described for reducing the basilar invagination, restoring craniospinal alignment, and establishing fixation of the atlantoaxial joint.[9] The advance in osteosynthesis now allows for a solid instrumented fusion. Plates and screws at the occiput combined with C1 lateral masses and C2 pedicular screws, transarticular C1–C2 screws, or translaminar C2 screws and any combination thereof with autograft or mixed allograft fusion have achieved solid fusion.

7. *After posterior fusion, what is the natural history of the basilar invagination?*
   - There is evidence in the literature suggesting the regression of the soft pannus after internal fusion.[10]
   - A high rate of postoperative morbidity (42%, $n = 55$), both early and 6 months mortality (13 and 25%, respectively) are reported for nonambulatory patients.[11]
   - Significant postoperative motor recovery was observed for a patient with a posterior atlantodental interval of > 14 mm.[12]

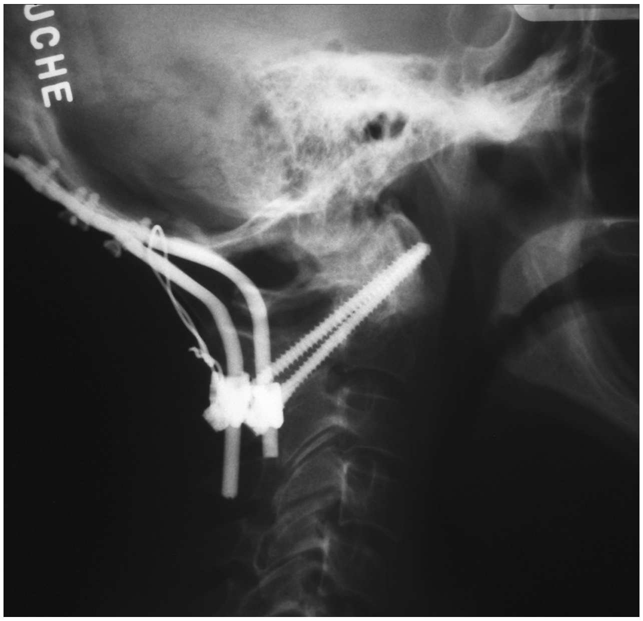

**Fig. 104.2** Cervical spine lateral radiograph. An instrumented occipitocervical fusion with occipital screws and transarticular atlantoaxial screws is visible.

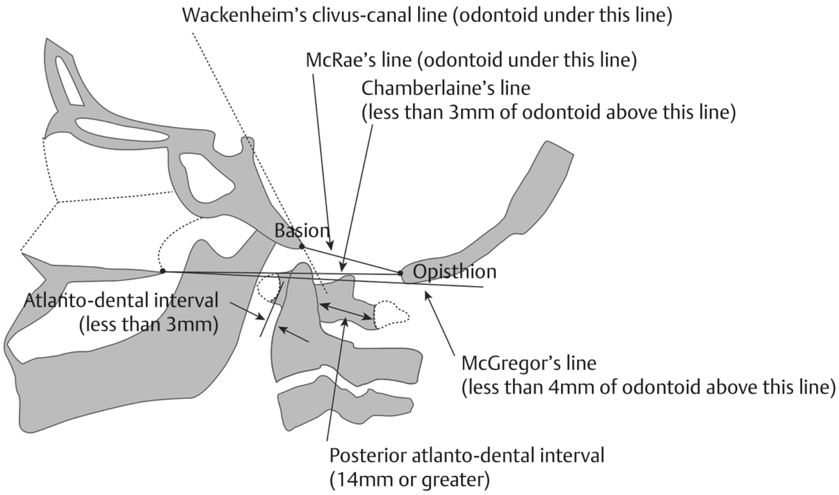
Wackenheim's clivus-canal line (odontoid under this line)
McRae's line (odontoid under this line)
Chamberlaine's line
(less than 3mm of odontoid above this line)
Basion
Opisthion
Atlanto-dental interval
(less than 3mm)
McGregor's line
(less than 4mm of odontoid above this line)
Posterior atlanto-dental interval
(14mm or greater)

**Fig. 104.3** Adult craniometric lines (and normal values) in lateral view.

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# Case 105 Central Cord Syndrome/Cord Contusion

*Daniel S. Ikeda, Andrew Shaw, and H. Francis Farhadi*

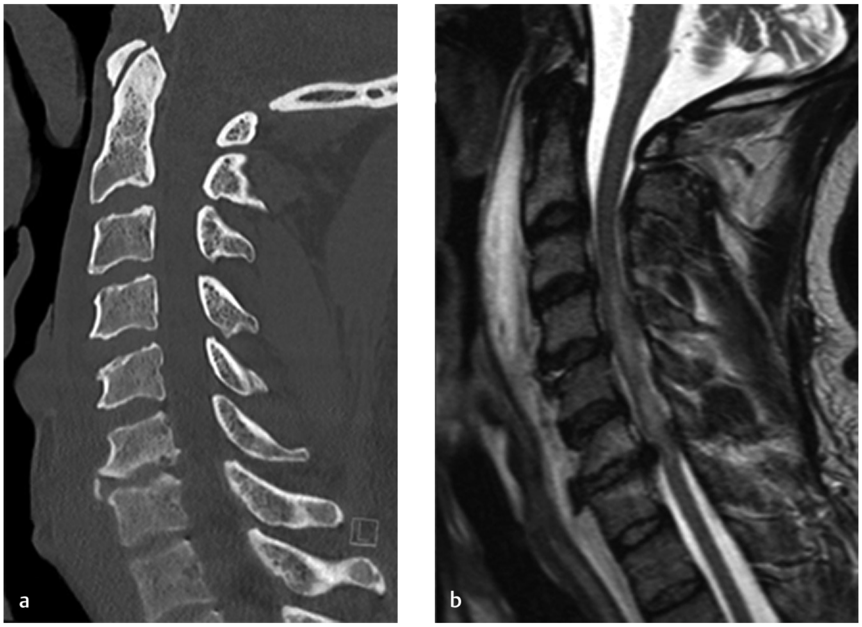

**Fig. 105.1** A sagittal CT scan of the cervical spine demonstrates multilevel degenerative changes with no fracture or malalignment (**a**). A sagittal MR T2 demonstrates significant prevertebral edema, intramedullary spinal cord edema, increased T2 signal in the C4–C5 disc space concerning for ligamentous disruption, and preexisting multilevel degenerative stenosis (**b**).

## Clinical Presentation

- A 59-year-old man presents to the emergency department after a fall where he struck his head on a sign. He has been nonambulatory since the fall and has minimal neck pain. He also complains of severe burning pain in his arms.
- On physical exam, he is awake and oriented. He has a laceration on his forehead. He demonstrates 3/5 strength (Medical Research Council [MRC] motor scale) in his deltoids and biceps, and 2/5 strength in his triceps and distal upper extremity (UE) muscles bilaterally. He has 4/5 strength in his lower extremities (LEs) throughout all motor groups tested. In addition, he demonstrates significant allodynia to light touch over his bilateral UEs, but otherwise has preserved sensation.
- A CT scan of the head is negative and CT of the cervical spine is negative for fractures or misalignment (▶**Fig. 105.1**a). A cervical MRI is then obtained (▶**Fig. 105.1**b).

## Questions

1. What is the diagnosis and what is the typical mechanism of this injury?
2. What is the American Spinal Injury Association (ASIA) impairment scale for this injury?
3. Describe the CT and MRI findings.
4. What is the significance of increased T2 signal within the spinal cord on MRI?
5. What is your initial management?
6. What are the indications for surgery and potential surgical strategies?
7. If indicated, what is the optimal timing for surgery?
8. What are potential variables that predict prognosis after such an injury?
9. Another patient presents with similar stenosis found incidentally. How would you counsel the patient regarding the potential risks of developing such an injury in the future and indications for a “prophylactic” surgery?

## Answers

1. *What is the diagnosis and what is the typical mechanism of this injury?*
   - This is the classic presentation for central cord syndrome (CCS)
     - Patients typically have a symmetric and disproportionately greater motor deficit in the UEs as compared to the LEs, with more pronounced weakness in the hands than the proximal UEs.
     - While there are no absolute criteria for diagnosis, some experts have suggested the requirement of a difference of at least 10 motor score points (according to the International Standards for Neurological Classification of Spinal Cord Injury) between the UEs and LEs.[1–3]
     - As many as 22% of patients experience hyperpathia or allodynia, which often subsides within 2 to 3 weeks.[4]
     - The majority of patients are middle aged and older men.
   - This is the most common incomplete spinal cord injury (SCI) syndrome with an approximate 3% inpatient mortality rate.[5]
   - The mechanism is most typically an acute hyperextension injury that results in disproportionate direct injury to the *lateral corticospinal tracts.*
     - Patients with preexisting stenosis often have anterior canal bony degenerative changes with posterior redundant ligamentum flavum (LF). Some patients may have congenital stenosis or preexisting instability.
     - As in this patient, there may be signs of trauma to the face or head incurred during the injury.
   - The reason why the UE findings are generally more pronounced than the LE findings is likely related to the fact that the corticospinal tract serves mainly the distal limb musculature, therefore its injury results in a functionally more pronounced deficit in the hands.[6] It is also related to the fact that fibers responsible for LE motor functions are located in the most peripheral part of the cord whereas fibers controlling the UE motor functions are more centrally located.
2. *What is the American Spinal Injury Association (ASIA) impairment scale for this injury?*
   - The patient presents with an ASIA grade D SCI with a C4 level.
     - He has preserved sensation with at least 3/5 strength in more than half of the muscle groups tested below the level of the injury.
   - His admission ASIA total motor score is 64 (UE score 24 and LE score 40).
3. *Describe the CT and MRI findings.*
   - The CT shows no fracture or malalignment, with multiple levels of degenerative changes.
   - The MRI T2 sagittal slice demonstrates extensive prevertebral edema, increased signal within the C4–C5 interspace concerning for anterior longitudinal ligament disruption, degenerative central canal stenosis and cord compression, and generalized spinal cord edema consistent with SCI.
4. *What is the significance of increased T2 signal within the spinal cord on MRI?*
   - Intramedullary increased T2 signal is an indication of contusional SCI in this setting.
     - Acute compression from LF buckling with hyperextension causes injury to the cord and resultant edema.
   - Postmortem pathological examination in CCS has demonstrated axonal injury without hemorrhage and intact central gray matter in patients with increased intramedullary T2 signal.[7]
5. *What is your initial management?*
   - Evaluation according to the Trauma Team and Advanced Trauma Life Support protocol to identify other potential systemic injuries
   - Spinal precautions and rigid cervical orthosis given the potential of ligamentous instability
   - Admission to intensive care unit with frequent neurological assessments
   - Maintenance of a mean arterial pressure of 85 mm Hg for up to 7 days post injury in patients without a significant cardiac history is a Level III recommendation for acute traumatic SCI.[8]
   - While controversial, the most recent data suggests that there is no evidence for steroid administration in SCI (Level 1 evidence), and, in fact, administering steroids to these patients may well have deleterious complications.
6. *What are the indications for surgery and potential surgical strategies?*
   - Patients should undergo decompression/stabilization surgery if they have ongoing cord compression at the level(s) that correlate with the neurological deficit, continued deterioration of function, or instability of the spine.
   - Similar to other pathologies of the cervical spine, surgery should be targeted to the area(s) of noted cord compression.
     - For patients with ventral compression, anterior cervical decompression and fusion should be considered.
     - For patients with significant dorsal compression from ligamentous buckling, a posterior cervical decompression with arthrodesis should be considered (see ▶**Fig. 105.2**).
     - Rarely, patients will be unstable or have circumferential stenosis potentially requiring a combined anterior and posterior surgery.

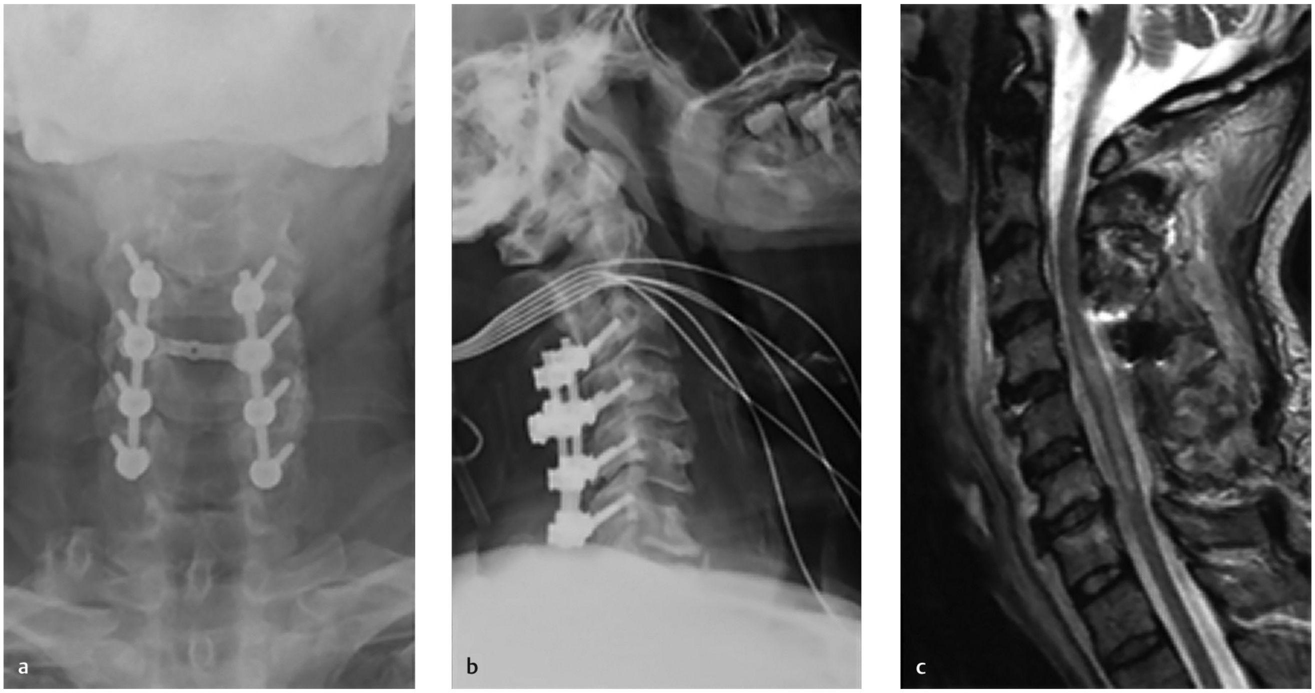

**Fig. 105.2** Anteroposterior and lateral postoperative cervical X-rays demonstrate laminectomies and satisfactory placement of posterolateral lateral mass fixation (**a, b**). A postoperative MRI demonstrates adequate spinal cord decompression and evidence of spinal cord expansion with intramedullary edema (**c**).

## Answers (*continued*)

7. *If indicated, what is the optimal timing for surgery?*
   - Classic teaching and practice has been to wait and observe the patient to allow the patient to improve without surgical intervention.[9]
     - Earlier CCS series pointed to a tendency to deteriorate after acute surgery.[10]
   - In contrast, a recent trend in care has demonstrated that early surgery is not only safe but also may be associated with enhanced improvement in function.[5]
     - The Surgical Timing in Acute Spinal Cord Injury Study (STASCIS) suggested a greater chance for ASIA score improvement in patients with acute SCI (including CCS) operated on within 24 hours.[11]
     - If possible, early surgery within the first 24 hours after injury should be considered, provided it can be done safely. Otherwise, surgery should ideally be performed within a few weeks. *The patient underwent a wide decompressive laminectomy and posterior C3–C6 arthrodesis with fusion. For five days after admission, he was kept in an intensive care unit for blood pressure management to maintain a mean arterial pressure of at least 85 mm Hg. He was discharged to inpatient rehabilitation on postoperative day 7. At 4-weeks follow-up, he was found to have improved strength in the left UE and he was able to ambulate independently with a cane.*
8. *What are potential variables that predict prognosis after such an injury?*
   - Patients with an admission ASIA grade of D had significantly greater functional independence than those presenting with an ASIA grade C examination.[12]
   - Perhaps counterintuitively, a smaller midsagittal central canal diameter was associated with improved neurological outcome.
   - Younger patient age and greater length of intrinsic spinal cord MR T2 signal were associated with increased dysesthetic pain at 6-month follow-up.
9. *Another patient presents with similar stenosis found incidentally. How would you counsel the patient regarding the potential risks of developing such an injury in the future and indications for a “prophylactic” surgery?*
   - In a small cohort study, 55 asymptomatic or minimally symptomatic patients with a mean midsagittal cervical canal diameter of 6.1 mm were followed for an average of 2.3 years without surgery.[13]
     - Despite 10 traumatic events in the follow-up period, there were no SCIs.
   - While the majority of patients who develop CCS have preexisting cervical stenosis, there is no consensus regarding prophylactic surgery. Asymptomatic patients should be counseled that the incidence of acute neurological deterioration after trauma is not known, but is likely very low.

## Suggested Readings

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# Case 106 Lower Cervical Fracture Dislocation

*Christopher Evan Stewart, Joseph A. Shehadi, and Brian Seaman*

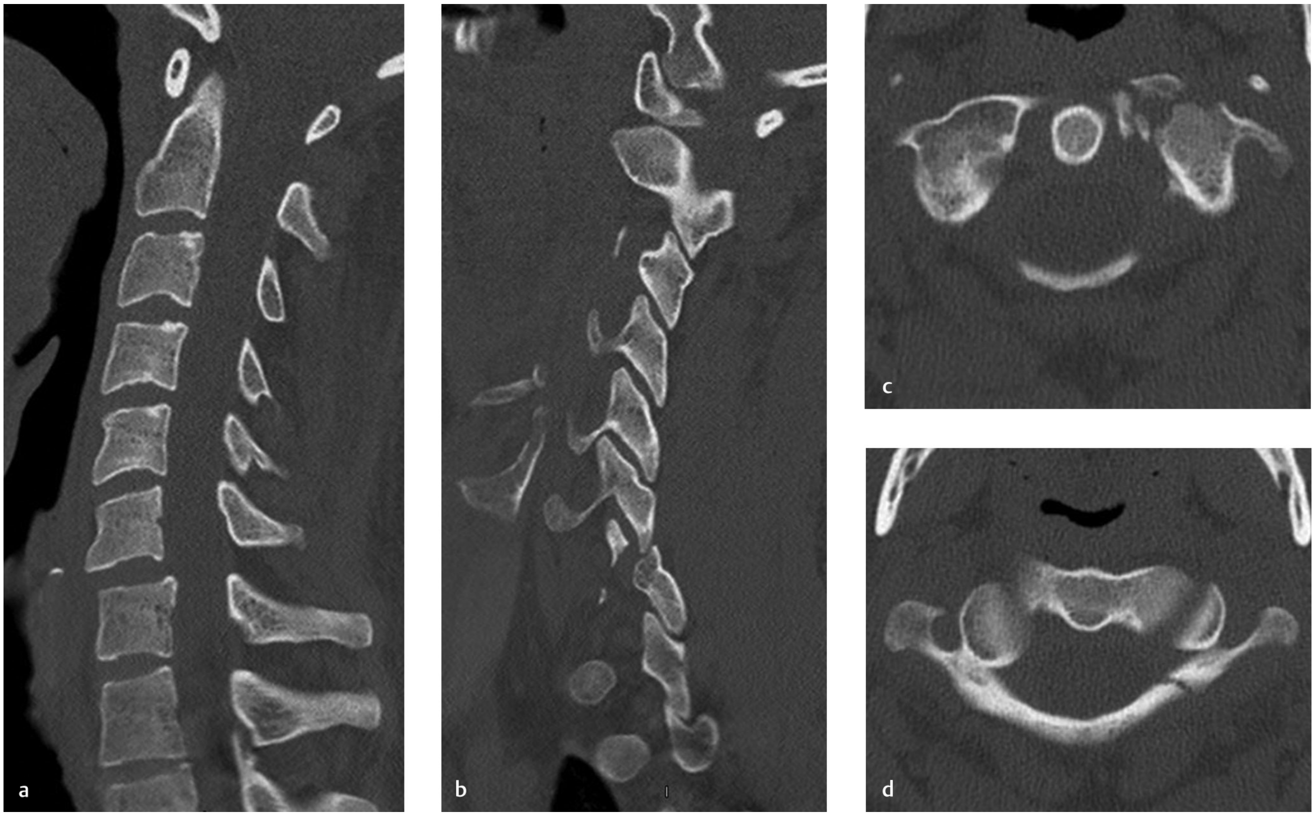

**Fig. 106.1** CT scan of the cervical spine with (**a**) midsagittal view, (**b**) parasagittal view through the right-sided facet joints, and (**c, d**) axial views of C1.

## Clinical Presentation

- A 36-year-old police officer with no past medical history was involved in a motor vehicle collision. Since the accident he reports pain and numbness in his right hand and wrist.
- On neurologic examination he had Glasgow Coma Score (GCS) of 15, cranial nerves II to XII were intact, he was found to have 5/5 in bilateral deltoids, biceps, triceps, wrist extensors, interossei, iliopsoas, quadriceps, dorsiflexion, extensor hallucis longus, and plantar flexion. He suffers of decreased sensation to light touch and pinprick over his right second and third digits. He had no long tract signs and reflexes were normoactive.
- CT of the cervical spine was obtained, which is illustrated in ▶**Fig. 106.1**.

## Questions

1. Interpret the CT scan of the cervical spine.
2. What is your initial management for this patient?
3. Are there any additional studies you would order?

   An MRI of the cervical spine was performed which was negative for any compressive spinal cord lesion. There was high short tau inversion recovery (STIR) signal intensity in the region of C6–C7 facets, as well as the interspinous and intraspinous ligaments. An MRA of the cervical spine was performed which revealed no evidence of blunt cerebrovascular injury.
4. Is there a role for closed reduction in cervical fracture dislocations? Should traction be used in this patient?

## Questions (*continued*)

5. Describe the novel classification systems for subaxial cervical spine injuries.
6. Calculate the Subaxial Injury Classification and Severity Scale (SLIC) score for this patient.

You decide that surgical intervention is indicated for this patient. This would allow for safe and expedient mobilization, as well as preservation of the neural elements.

7. Discuss your goals of surgery and your proposed surgical plan.
8. What recommendations would you have for the anesthesia staff prior to surgical intervention?
9. List the potential risks associated with anterior approaches to the cervical spinal column.
10. Discuss how you would proceed if you encountered an intraoperative dural tear during your anterior approach.
11. During placement of one of the left lateral mass screws you encounter substantial arterial bleeding, which you presume is the vertebral artery. Describe how you would proceed if a vertebral artery were injured intraoperatively.

The patient successfully underwent a combined anterior and posterior cervical fusion. Antiplatelet therapy was initiated to treat the vertebral artery dissection. Gabapentin was initiated for the treatment of the patient's neuropathic sensory complaints.

12. Detail an outpatient follow-up plan for this patient.
13. Discuss the potential delayed surgical complications that may occur after a cervical instrumented fusion.

## Answers

1. *Interpret the CT scan of the cervical spine.*
   - Midsagittal views of a grade 1 anterolisthesis of C6–C7. Right parasagittal views show a perched C6–C7 facet and displaced fracture of the superior articulating process of the C7 facet.
   - Axial views of C1 show a left posterior arch fracture and comminuted fracture affecting the left anterior arch and lateral mass.
2. *What is your initial management for this patient?*
   - The management of airway, breathing, and circulatory stability should be maintained according to Advanced Trauma Life Support protocol.
   - Spinal precautions and cervical spine immobilization should be maintained and a rigid cervical orthosis should be implemented.[1]
   - A thorough neurologic assessment should be completed with continued monitoring of any deficits.
3. *Are there any additional studies you would order?*
   - MRI is helpful in this patient to determine whether ligamentous injury is present. Injury to the anterior longitudinal ligament (ALL), posterior longitudinal ligament (PLL), posterior interspinous and supraspinous ligaments, facet capsules, and disc spaces can be appreciated. It is critical to identify injury to these structures because this affects the surgical approach.
   - In addition, MRI is the study of choice to evaluate the neural elements if neurologic signs or symptoms are present.
   - MR angiography or CT angiography is recommended in these individuals to assess for blunt cerebrovascular injury, given the patient's distracting mechanism, bony subluxation, and lateralizing neurologic symptoms.[2]
4. *Is there a role for closed reduction in cervical fracture dislocations? Should traction be used in this patient?*
   - Early closed reduction of cervical spinal fracture dislocations with craniocervical traction for the restoration of anatomical alignment of the cervical spine in awake patients is recommended.[3]
   - Closed reduction is contraindicated in this particular patient due to the existence of an additional rostral injury.[3]
5. *Describe the novel classification systems for subaxial cervical spine injuries.*
   - The Subaxial Injury Classification and Severity scale is a novel metric which considers the morphologic appearance, evidence of ligamentous injury, and neurologic status of the patient to help guide surgical decision-making. In comparison to prior classification systems, it provides a more detailed description of the patient's injury and a favorable interrater reliability (**Table 106.1**).[4,5]
   - The Cervical Spine Injury Severity Score (CSISS) was put forward by Anderson et al in 2007.[6] It seeks to classify discoligamentous injury by the degree of skeletal or osseous displacement on CT scan. The cervical spine motion segment is divided into four columns: 1—vertebral body, including the ALL, PLL, and annulus; 2—the right facet and joint capsule; 3—the left facet and capsule; 4—the laminae including the spinous processes, pedicles, and interspinous and intraspinous ligaments. Each column is scored 0 to 5 in correlation to the skeletal or fracture line separation (0–5 mm). The individual column scores are summated for a final score of 0 to 20. Anderson et al recommended surgical

## Answers (*continued*)

fixation for scores of 7 or higher.[5] (see ▶**Fig. 106.2** for illustration of a case requiring fixation).

6. *Calculate the Subaxial Injury Classification and Severity Scale (SLIC) score for this patient.*
   - Total score: 6. Scores > 5 are considered in need of surgical fixation.[4]
7. *Discuss your goals of surgery and your proposed surgical plan.*
   - Closed reduction is not an option due to the patient's C1 fractures, therefore fracture reduction must be done surgically. Surgical reduction can be performed via either an anterior or posterior approach.
   - After normal alignment has been returned, the fracture should be stabilized. This patient's SLIC score was 6 placing him into the operative category. Internal fixation and fusion by either anterior or posterior procedures have been shown to be effective. The need for decompression may also change which approach is used.[7]
   - This patient underwent anterior discectomy of C6–C7 followed by intraoperative reduction under fluoroscopic guidance. A C6–C7 interbody was placed and secured with anterior platting. He was then approached posteriorly where C6–C7 lateral mass screws and arthrodesis were performed along with a right C6–C7 foraminotomy, partial facetectomy, and retrieval of the superior C7 fracture fragment.
8. *What recommendations would you have for the anesthesia staff prior to surgical intervention?*
   - Videolaryngoscopy or awake fiberoptic intubation technique should be considered, given the cervical spine instability.
   - Prevention of intraoperative hypotension (often during induction) and possible ischemia is critical in cases with spinal cord injury or blunt cerebrovascular injury.
9. *List the potential risks associated with anterior approaches to the cervical spinal column.*
   - The most common complications of anterior approaches to the cervical spine include recurrent laryngeal nerve palsy with subsequent hoarseness, dysphagia, and odynophagia.[8] Complete paralysis of the vocal cords is rare and is estimated to be around 0.5%.[8]
   - Other less common complications include hematoma, large vessel arterial or venous injury/thrombosis, esophageal injury, dural sac manipulation with neurologic injury, dural tear, breathing difficulties or delayed extubation secondary to soft tissue swelling, traction C5 palsy, peripheral and cranial nerve injury (hypoglossal), vertebral artery injury, Horner's syndrome from sympathetic chain injury, and thoracic duct injury.[9]
10. *Discuss how you would proceed if you encountered an intraoperative dural tear during your anterior approach.*
    - One can attempt a primary repair utilizing microsurgical techniques.
    - Frequently, Gelfoam (Pfizer Pharmaceuticals, New York, NY) or collagen matrix (preferably inlay) and fibrin glue or hydrogel sealant can be placed over the defect, with care not to compress or manipulate the cord.
    - Lumbar drainage can be used as an adjunctive measure.[9]
11. *During placement of one of the left lateral mass screws you encounter substantial arterial bleeding, which you presume is the vertebral artery. Describe how you would proceed if a vertebral artery were injured intraoperatively.*
    - If the vertebral artery is injured from a posterior approach, then direct repair is not feasible. The screw should be kept in and intraoperative or immediate postoperative angiography is then performed.
    - Endovascular obliteration is a good option for the management of a pseudoaneurysm after vertebral artery injury as long as the injured vertebral artery is not dominant.[10]
12. *Detail an outpatient follow-up plan for this patient.*
    - Rigid cervical orthosis should be maintained postoperatively secondary to his C1 fractures.
    - The patient should initially follow up in the first 2 weeks for wound observation and preservation of spinal alignment.
    - Follow-up appointment in 12 weeks should include plain cervical radiographs. Flexion and extension views can be performed to confirm spinal stability.
    - A hard collar should be maintained for at least 12 weeks. At the time of radiographic fusion, remove the collar and begin isometric exercises and physical therapy.
13. *Discuss the potential delayed surgical complications that may occur after a cervical instrumented fusion.*
    - Delayed complications following cervical fusion include pseudoarthrosis, hardware failure, and postsurgical kyphosis.
    - Pseudoarthrosis is a known cause of persistent neck pain and radiculopathy following anterior cervical discectomy and fusion.[11] Pseudoarthrosis rates increase with multilevel constructs and may be as high as 50% for three-level fusions.[8] In patients who undergo anterior cervical surgery with resultant symptomatic pseudoarthrosis, a posterior fusion may be more effective than anterior revision.[12] In addition, anterior cervical plating has reduced the incidence of pseudoarthrosis.

**Table 106.1** Subaxial injury classification and severity scale

| Morphology | Points |
|---|---|
| No abnormality | 0 |
| Compression | 1 |
| Burst | + 1 = 2 |
| Distraction (facet perch, hyperextension) | 3 |
| Rotation/translation (facet dislocation, unstable teardrop, or advanced stage flexion compression injury) | 4 |
| **Discoligamentous complex** | |
| Intact | 0 |
| Indeterminate (isolated interspinous widening. MRI signal change only) | 1 |
| Disrupted (widening of disc space, facet perch or dislocation) | 2 |
| **Neurologic status** | |
| Intact | 0 |
| Root injury | 1 |
| Complete cord injury | 2 |
| Incomplete cord injury | 3 |
| Continuous cord compression in setting of neurologic deficit | + 1 = 1 |

Source: Vaccaro et al[4]

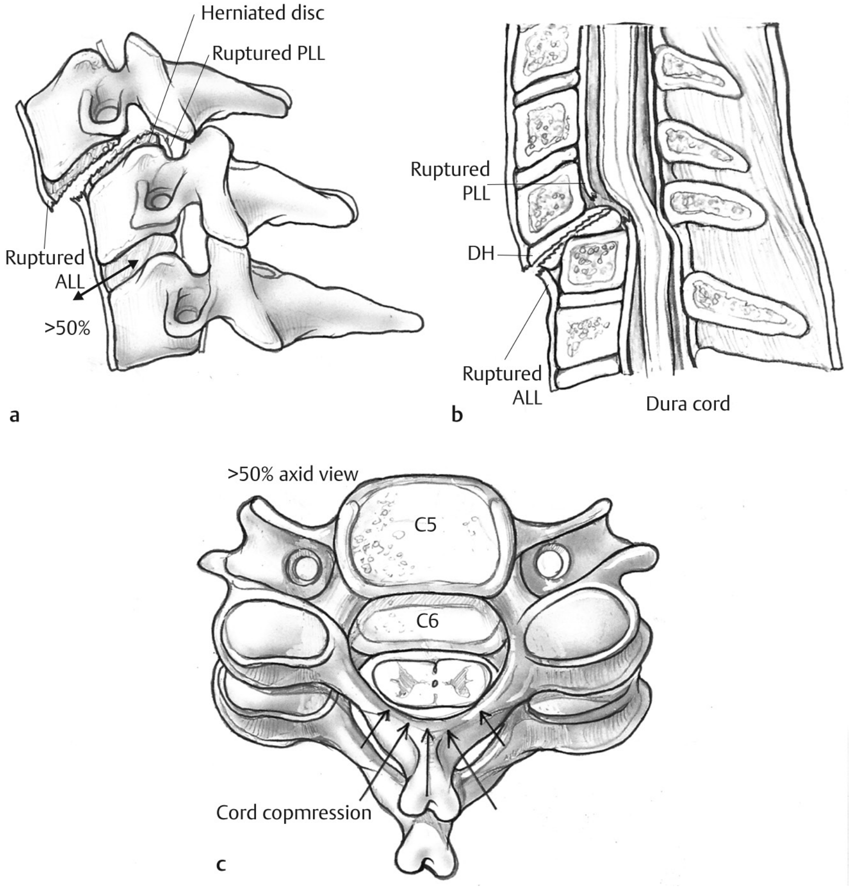

**Fig. 106.2** (**a**) Artist's rendering of jumped facets with disc disruption. (**b**) Sagittal section showing spinal cord injury at the level of the disc disruption/herniation with impingement and severe stenosis. (**c**) Axial view with subluxation of C5 over C6 vertebrae with canal narrowing and cord impingement. (Reproduced with permission from Neurosurgery Tricks of the Trade—Spine and Peripheral Nerve, Nader R., et al., New York: Thieme; 2013)

## Answers (*continued*)

- Hardware failure or screw loosening typically can be seen at the caudal end of a multilevel construct.[13] Graft displacement can be seen in 2 to 8% of cases.
- These complications may be avoided by maximizing screw purchase, preparing a well-fitting graft under compression, and possibly by the use of dynamic anterior cervical plating.[8,13]
- Postsurgical kyphosis may result in patients suffering significant two-column injuries who undergo anterior cervical surgery alone. Adjunctive-instrumented posterior fusion can reduce the incidence of this complication.
- Adjacent segment disease can occur above or below the fusion.

## Suggested Readings

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# Case 107 Thoracic Compression Fracture without Neurological Deficit

*Jorge E. Alvernia, Jorge E. Isaza, Eddie Perkins, and Edgar Gerardo Ordonez-Rubiano*

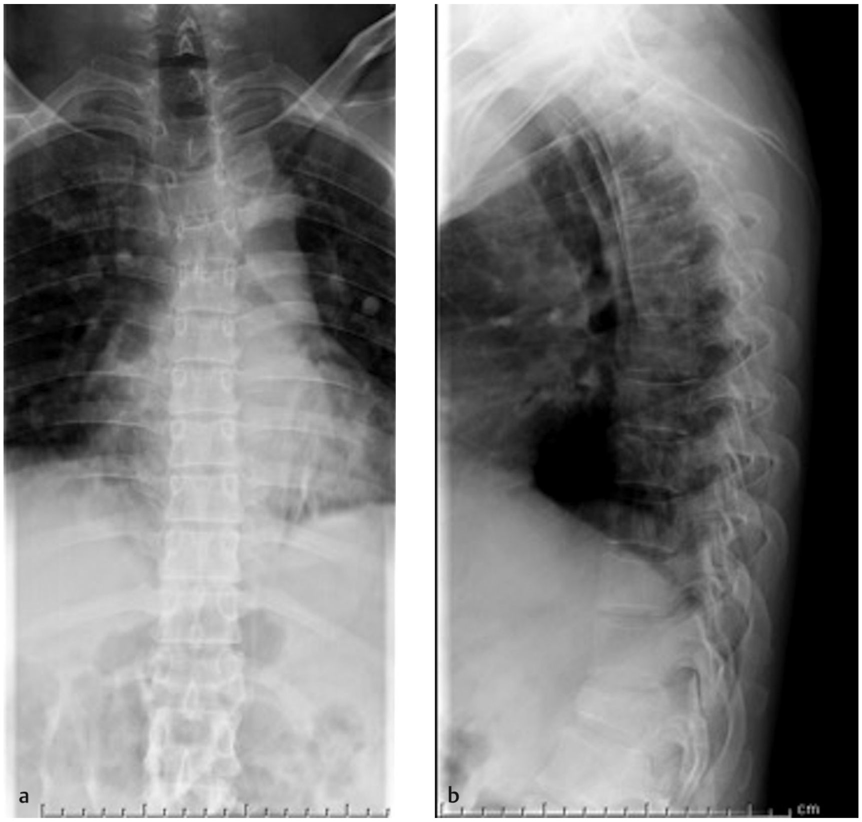

**Fig. 107.1** (**a**) Anteroposterior and (**b**) lateral X-rays of the thoracolumbar spine.

## Clinical Presentation

- A 39-year-old man who fell from a bridge presents to the ER complaining of upper back pain.
- Past medical history includes alcohol, cannabinoids, and cocaine use.
- Upper back midline point tenderness is present. Thoracic and lumbar range of motion is limited because of severe pain. No neurological deficits are present.
- Thoracic spine X-rays (▶**Fig. 107.1**a, b), thoracic spine CT (▶**Fig. 107.2**a) and thoracic spine MRI (▶**Fig. 107.2**b) were obtained.

## Questions

1. Describe the images presented in ▶**Fig. 107.1** and ▶**Fig. 107.2**a, b.
2. What kind of injury mechanism led to this fracture?
3. Does the term unstable according the Denis three-column classification automatically means that a surgical intervention is necessary?
4. Describe the importance of spine angulation for the treatment of this kind of fractures.
5. Describe the Thoracolumbar Injury Classification and Severity Scale (TLICS) and the reason this classification is becoming the most accepted.
6. Describe the components of the posterior ligamentous complex (PLC).
7. What CT findings are suggestive of PLC involvement?
8. What MRI findings are suggestive of PCL involvement?
9. What treatment do you recommend in this patient according to the TLCIS classification?
10. What is the recommended time frame to perform surgery in this case?
11. What are the goals of surgery?
12. What kind of surgical approach would you recommend?

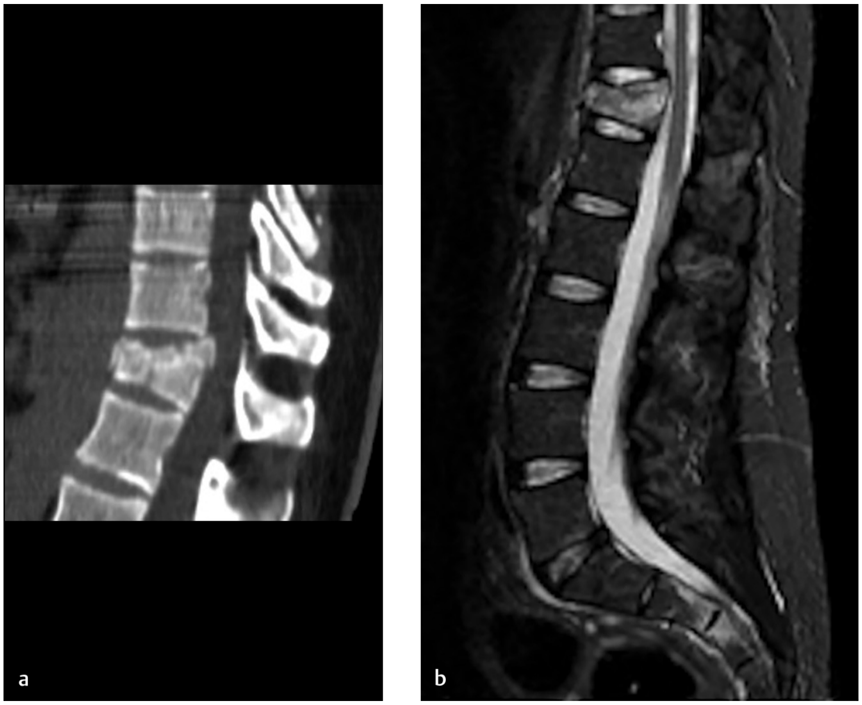

**Fig. 107.2** (**a**) CT scan reconstructed sagittal sections of the thoracolumbar spine, bone windows. (**b**) MRI T2-weighted image, midsagittal section of the thoracolumbar spine.

## Answers

1. *Describe the images presented in* ▶**Fig. 107.1** *and* ▶**Fig. 107.2**a, b.
   - Thoracic X-rays depict a T12 burst fracture. Thoracic spine CT depict a T12 burst fracture with 50% decrease of the vertebral high and retropulsion into the canal resulting in about 20% decrease of the spinal canal diameter. An approximate 25 degrees of kyphotic angulation is present as well.
   - Thoracic spine MRI did not show T2 signal increase with regard to the PLC; however, the thoracic CT shows widening of the interspinous space suggestive of posterior ligament injury.
2. *What kind of injury mechanism led to this fracture?*
   - Burst fractures usually are the result of compression with severe axial loading.
3. *Does the term unstable according the Denis three-column classification automatically means that a surgical intervention is necessary?*
   - The term “spine instability” in the original three-column classification does not necessarily mean “need for surgical intervention,” however, should the patient have presented with neurological deficits, then he would have been a potential surgical candidate.[1]
4. *Describe the importance of spine angulation for the treatment of this kind of fractures.*
   - Additional parameters have been defined in order to help in the surgical decision-making. Twenty or more degrees of spine kyphotic angulation and more than 3.5 mm vertebral listhesis are suggestive of ligament injury, both of them in favor of a surgical intervention.
5. *Describe the thoracolumbar Injury Classification and Severity Scale (TLICS) and the reason this classification is becoming the most accepted.*
   - Because of the prior classifications’ failure to systematically take into account the neurological status of the patient and the indications for MRI to determine the integrity of the PLC in 2005 the Trauma Study Group introduced the TLICS in order to facilitate the surgical decision-making.[2,3]
   - The TLICS consists of three independent parameters:
     - Injury morphology
     - Integrity of the PLC
     - Neurological status
   - A parameter can be scored from 0 to 4 points and the total score is the sum of these parameters with a maximum of 10 points.
   - The total score predicts the need for surgery as is shown in the ▶**Table 107.1**. A total of 5 or more points indicates the need for surgical treatment.[4]
6. *Describe the components of the posterior ligamentous complex (PLC).*
   - The PLC is composed of the supraspinous ligaments, interspinous ligaments, articular facet capsules, and ligament flava.
7. *What CT findings are suggestive of PLC involvement?*
   - Widening of the interspinous space

**Table 107.1** Thoracolumbar Injury Classification and Severity Scale (TLCIS)

| TLCIS | | | | |
|---|---|---|---|---|
| 1 | Morphology<br>Immediate stability | Compression<br>Burst<br>Translation/rotation<br>Distraction | 1<br>2<br>3<br>4 | Radiographs<br>CT |
| 2 | Integrity of PLC<br>Long-term stability | Intact<br>Suspected<br>Injured | 0<br>2<br>3 | MRI |
| 3 | Neurological status | Intact<br>Nerve root<br>Complete cord<br>Incomplete cord<br>Cauda equina | 0<br>2<br>2<br>3<br>3 | Physical examination |
| 4 | Predicts | Need for surgery | 1–3<br>4<br>> 4 | Nonsurgical<br>Surgeon's choice<br>Surgical |

## Answers (*continued*)

- Avulsion fractures or transverse fractures of spinous processes or articular facets
- Widening or dislocation of facet joints
- Vertebral body translation or rotation

8. *What MRI findings are suggestive of PCL involvement?*
   - Loss of normal low signal intensity of the ligaments flava or supraspinous ligaments on T1 and T2
   - High signal intensity of the interspinous ligaments or along the facet joints on T2 spectral presaturation with inversion recovery (SPIR) or short tau inversion recovery (STIR) sequences[2,3]
9. *What treatment do you recommend in this patient according to the TLCIS classification?*
   - According to the TLCIS classification, the total score of this patient will be 4 points:
     - Morphology: burst (2 points)
     - Integrity of PLC: although the MRI did not show T2 changes in relationship to the PLC, the widening of the interspinous space distance and the kyphotic angulation of 25% degree raise the suspicion for PLC involvement[1]
     - Neurological status: intact (0)
   - Based on the recommendations of this classification, the recommendation in this particular case is surgeon's choice, i.e., either to treat conservatively with a thoracolumbosacral orthotic (TLSO) brace[4] or to offer surgery.
   - If brace is recommended in this case, follow-up imaging is strongly recommended.[5,6]
   - On the other hand, if surgery is performed, a posterior instrumentation with transpedicular screws and rods two or three levels above and two levels below is an option[7].
10. *What is the recommended time frame to perform surgery in this case?*
    - Early intervention (8–24 hours after the accident) provided that patient's general conditions allow the surgical intervention and there are no other contraindications (medical, system or other) to surgical procedures.
11. *What are the goals of surgery?*
    - Relieve pressure from the spinal cord
    - Achieve spinal stability and fusion
12. *What kind of surgical approach would you recommend?*
    - An anterior or posterolateral approach is needed because of the retropulsion within the canal that has resulted in neurological deficit.
    - As to the steps to achieve stability and fusion, in the authors' opinion, a combined approached including a long posterior transpedicular fusion is preferred because of the location of the fracture (thoracolumbar junction), as was done in this case (▶ **Fig. 107.3**).
    - However, an anterior or anterolateral decompression and instrumentation through the same approach is another reasonable alternative.
    - Strut settling due to age and/or bone quality have been argued as reasons in favor of combined anterior and posterior fusion.[8–10]

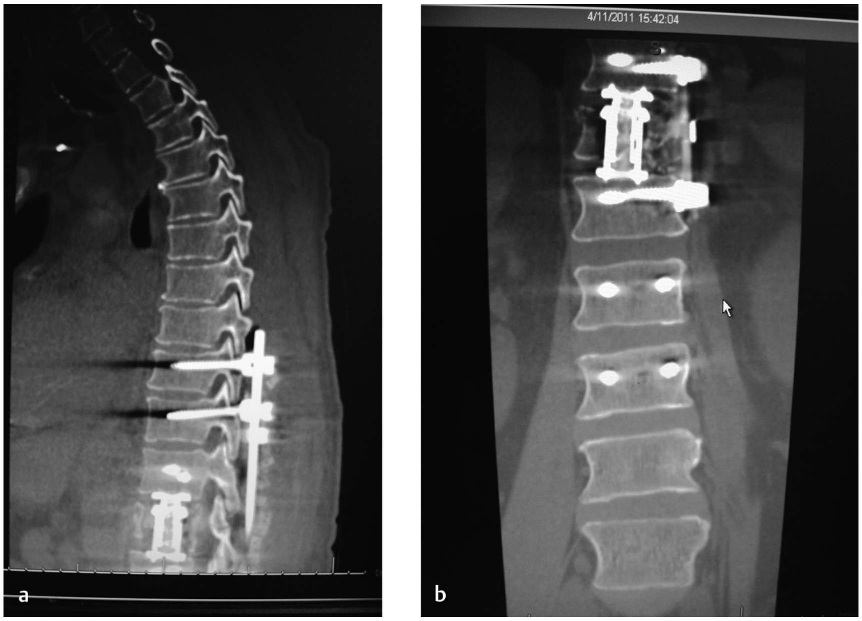

**Fig. 107.3** (**a, b**) A decision to perform a two-staged 360 degrees fusion was made. An anterior approach including a A T2 corpectomy with placement of a expandable cage plus a Kaneda plate was performed first followed one week after by a posterior transpedicular screw fixation two levels above and three levels below the fracture because the location of the fracture was at the thoracolumbar junction.

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# Case 108 Thoracolumbar Fracture—Dislocation with Complete Spinal Cord Injury

*Evan S. Marlin, Daniel S. Ikeda, Andrew Shaw, and H. Francis Farhadi*

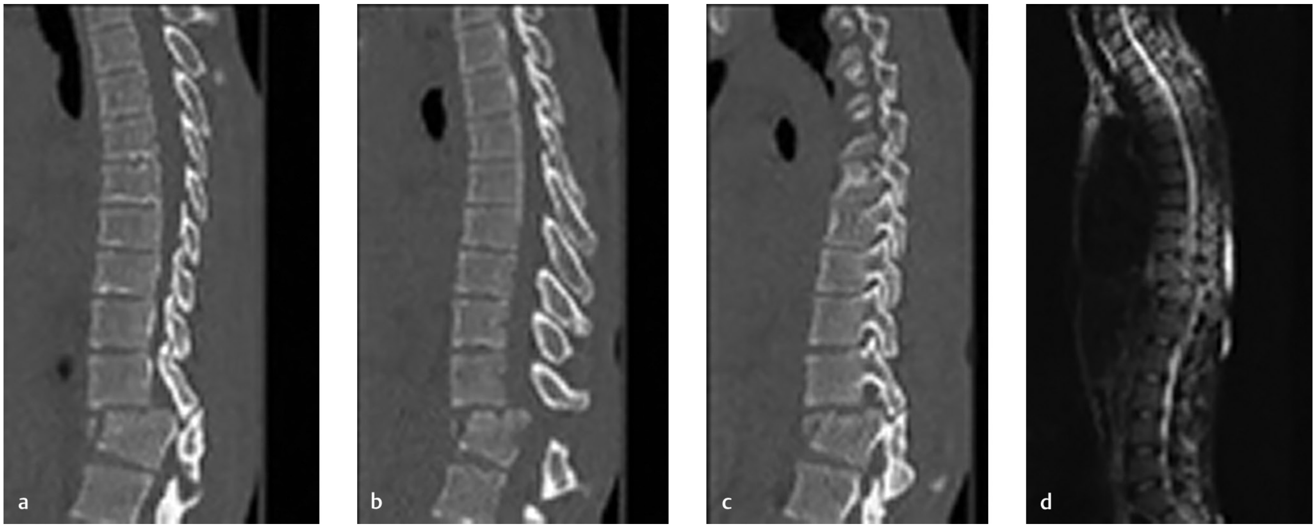

**Fig. 108.1** (**a–c**) CT scan reconstructed sagittal sections of the thoracolumbar spine, bone windows. (**d**) MRI T2-weighted image, midsagittal section of the thoracolumbar spine.

## Clinical Presentation

- A 28-year-old male presents to the emergency room as a level I trauma following a high-speed motor vehicle collision and ejection.
- An emergent cricothyroidotomy was performed in the field. Bilateral chest tubes are placed in the trauma bay for bilateral pneumothoraces.
- On secondary survey, he is noted to have a lower thoracic step-off. His eyes are open spontaneously; he localizes with his upper extremities but cannot move his lower extremities. He denies any sensation to pain or light touch from the lower thoracic levels and below. He has no rectal tone.

## Questions

1. What is his Glasgow Coma Score (GCS)? In determining his degree of injury, what are the critical neurologic reflexes to examine and what do they evaluate?
2. What is the American Spinal Cord Injury Association (ASIA) Impairment Scale (AIS)? What AIS grade is he?
3. Describe the imaging findings (▶**Fig. 108.1**).
4. What is the mechanism of injury? What bony elements and ligaments are injured?
5. What is the goal of surgery? What levels of fixation should be planned? How should reduction take place?
6. What other imaging should be completed at the time of his initial assessment?
7. What is the role of steroids in the treatment of acute spinal cord injury (SCI)?

## Answers

1. What is his Glasgow Coma Score (GCS)? In determining his degree of injury, what are the critical neurologic reflexes to examine and what do they evaluate?
   - E4 (eyes) V1-T (verbal – endotracheal tube in place) M5 (motor) = 10T
   - There are several spinal cord reflexes that can aid in localization and in determining the severity of neurological injury.
     - The *abdominal cutaneous reflex* involves the scratching of the upper and lower abdominal quadrants. The stimulated quadrant should flex and move the umbilicus in that direction. The upper abdominal reflex is useful for the T8–T9 level and lower abdominal reflex is indicative of T10–T12 function. This is a cortical reflex and its presence is indicative of an incomplete SCI.
     - The *cremasteric reflex* is another superficial reflex elicited by stroking the inner thigh to cause contraction of the cremaster muscle and elevation of the ipsilateral testicle. L1 and L2 sensory fibers from the genitofemoral nerve synapse in the cord and activate motor fibers of the genital branch of the genitofemoral nerve. Its presence is indicative of an incomplete SCI.
     - The *bulbocavernosus reflex* is a contraction of the anal sphincter in response to pinching the penis or in response to a Foley catheter tug. Its presence is indicative of an incomplete SCI. This reflex is mediated by S2–S4.
     - *Anal wink/anal–cutaneous reflex* is elicited by slight pinprick to the skin in the anal region resulting in involuntary anal contraction. It evaluates the most caudal sacral segments S2–S4.
2. *What is the American Spinal Cord Injury Association (ASIA) Impairment Scale (AIS)? What AIS grade is he?*
   - The AIS is a five-class scale that requires comprehensive assessment of motor and sensory impairment in patients with acute SCI (▶**Table 108.1**). It identifies the most rostral sensory and motor level of unimpaired function. It is based on the assessment of 28 bilateral dermatomes, motor testing of 10 muscle groups, and anal sensation/function.[1]
   - This patient has T12 ASIA A injury. On more detailed examination, the patient lacked sensation and motor function below his fracture. On rectal exam, there was no anal sensation, volitional tone, or anal wink. Sensation and motor function of the sacral segments should also be examined in delayed fashion to assess for complete injury after a period in which spinal shock should have resolved. The bulbocavernosus response is typically the first reflex to return.
3. *Describe the imaging findings (▶**Fig. 108.1**).*
   - The CT scan reveals a fracture-dislocation at T11–T12. There are fractures of bilateral T11–T12 facet joints with involvement of the bilateral T12 superior articular processes. There is significant anterolisthesis of T11 over T12 due to the bilateral jumped facet joints and an associated significant decrease in canal diameter. In addition, there is a compression fracture of the T12 vertebral body. The MRI reveals the anterior dislocation of T11 over T12 with ligamentous injury, severe cord compression, and associated edema.
4. *What is the mechanism of injury? What bony elements and ligaments are injured?*
   - This is a three-column injury associated with disruption of the anterior longitudinal ligament, posterior longitudinal ligament, and the posterior ligamentous complex.
   - Classically, thoracolumbar fracture-dislocations can occur as a result of flexion-rotation, flexion-distraction, and shear forces.[2,3]
     - Flexion-rotation injuries can cause anterior compression with disruption of the middle and posterior columns. There is often subluxation, increased interspinous distance, jumped facet joints, and minimal disruption to the posterior vertebral body.
     - Flexion-distraction injuries are similar to thoracolumbar Chance fractures and can cause subluxation with injury to the disc space.
     - Shear injuries often affect all three columns and are due to forces directed in a posterior-anterior direction. They often lead to fracturing of the posterior arch, facet joints, and spondylolisthesis with complete SCI. There may also be a rotational component to these injuries.
5. *What is the goal of surgery? What levels of fixation should be planned? How should reduction take place?*
   - Surgery should aim to decompress the spinal cord and correct the traumatic deformity. In this case, laminectomies and facetectomies at T11–T12

## Answers (*continued*)

will allow for reduction of the spondylolisthesis. Internal instrumented fixation will allow for earlier mobilization and decrease the incidence of progressive kyphotic deformity.
- The number levels of required fixation depends on the ability to stably reduce the fracture. Stable reduction can often be achieved through a posterior approach with two or three levels of fixation above and below the injury.
- If unable to reduce the spondylolisthesis, a corpectomy should be considered through either an anterior or dorsolateral approach.
- The load sharing score can aid in the determination of whether an injury can be addressed through a single posterior approach or if additional anterior stabilization is necessary.[4]
- In this case, three vertebral fixation levels above the dislocation acted as a cantilever to allow for reduction following complete T7–T8 laminectomies and facetectomies. These levels were reduced to the rod in sequential fashion (▶**Fig. 108.2**).

6. *What other imaging should be completed at the time of his initial assessment?*
   - Spinal injuries are present in up to 46% of traumas. Eighty percent of these injuries involve the thoracic and lumbar spine.[5,6]
   - Thoracic and thoracolumbar injuries are frequently associated with lung contusions, pneumothoraces, pleural effusions, and rib fractures.
   - Lumbar injuries are associated with viscus and pelvic injuries.
   - Patients presenting with injuries of the thoracolumbar spine from high-velocity or blunt-force injuries should be evaluated for intra-abdominal injury, pelvic injury, lung injury, and imaging of the cervical spine. Concomitant cervical spine injuries can occur in up to 25% of traumatic thoracolumbar fractures.[2] If a cervical fracture is present, angiographic imaging may also be warranted.[7]
   - MRI, while not imperative, aids in visualization of the spinal cord, supporting ligaments, and disc spaces. In addition, the identification of an epidural hematoma may alter surgical planning (timing and approach) and the extent of decompression performed.
7. *What is the role of steroids in the treatment of acute spinal cord injury (SCI)?*
   - The use of methylprednisolone (MPS) has been shown in model SCI systems to decrease spinal cord hemorrhage and enhance oligodendrocyte survival when started early.
   - However, clinical trial data have not shown improved neurological outcomes but rather an elevated risk of death and systemic complications including infection, sepsis, and wound breakdown.
   - As such, the routine use of high dose MPS is no longer recommended in trauma guidelines.[8,9]
   - The current recommendation is to treat all patients with SCI according to the local/regional protocol.
   - If steroids are recommended, they should be initiated within 8 hours of injury with the following steroid protocol: MPS 30 mg/kg bolus over 15 minutes and an infusion of MPS at 5.4 mg/kg/hour for 23 hours beginning 45 minutes after the bolus.

**Table 108.1** ASIA Impairment Scale (AIS)

| | |
|---|---|
| A: Complete | No sensory or motor function is preserved in the sacral segments S4–S5 |
| B: Sensory incomplete | Sensory but no motor function is preserved below the neurological level and includes the sacral segments S4–S5 (light touch, pin prick at S4–S5 or deep anal pressure), AND no motor function is preserved more than three levels below the motor level on either side of the body |
| C: Motor incomplete | Motor function is preserved below the neurological level and more than half of key muscle groups below the neurological level of injury (NLI) have a muscle grade less than 3 |
| D: Motor incomplete | Motor function is preserved below the neurological level and at least half of key muscle groups below the NLI have a muscle grade of 3 or greater |
| E: Normal | If sensation and motor function are graded as normal in all segments, and the patient had prior deficits, then the AIS grade is E. Someone without an initial SCI does not receive an AIS grade |

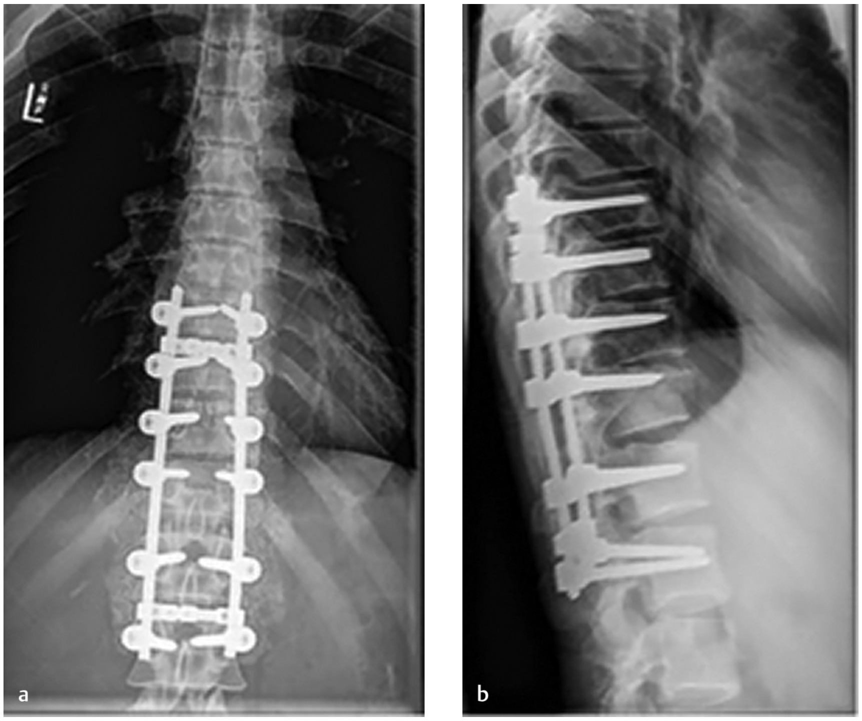

**Fig. 108.2** (**a**) Anteroposterior and (**b**) lateral X-rays of the thoracolumbar spine obtained postoperatively.

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# Case 109 Disc Disruption and Ligamentous Injury

*Anish Sen and Ibrahim Omeis*

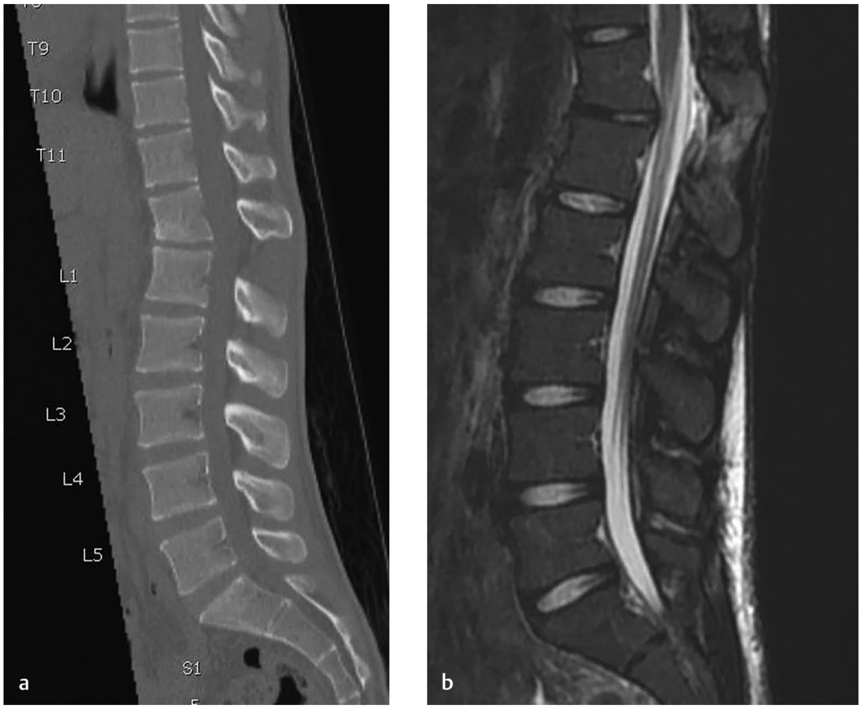

**Fig. 109.1** (**a**) CT scan reconstructed midsagittal section of the thoracolumbar spine, bone windows. (**b**) MRI T2-weighted image, midsagittal section of the thoracolumbar spine.

## Clinical Presentation

- A 22-year-old male presents to the emergency room after a motor vehicle accident, complaining of back pain.
- He denies any lower extremity pain or paresthesia and does not report bowel or bladder incontinence.
- On examination, he is hemodynamically stable and has no focal neurological deficits.
- A CT scan and MRI are obtained and are shown in ▶**Fig. 109.1**.

## Questions

1. Describe the CT and MRI findings (▶**Fig. 109.1**).
2. In addition to assessment of vital signs and a thorough neurological examination, what other components of the physical examination should be performed?
3. Describe the three-column classification system (of Denis) for the thoracolumbar spine.
4. Name the six ligaments of the thoracolumbar spine and define which movements they limit.
5. Describe the five relevant forces involved in structural damage to the thoracolumbar spine.
6. What are the advantages of early surgical stabilization compared to nonoperative management?
7. Describe the major nonoperative and operative approaches to treat traumatic disc disruption and ligamentous injury.
8. What anatomical features of the thoracolumbar junction make it susceptible to injury?

## Answers

1. *Describe the CT and MRI findings (▸* **Fig. 109.1***).*
   - The CT demonstrates a mild compression fracture of the T12 vertebral body with focal kyphosis at T12–L1.
   - There is also widening of the T12–L1 interspinous space, implying ligamentous injury.
   - The STIR-weighted MRI demonstrates hyperintensity in the disc space, as well as rupture of the posterior ligamentous complex. There is evident splaying of the spinous processes between T12 and L1.
2. *In addition to assessment of vital signs and a thorough neurological examination, what other components of the physical examination should be performed?*
   - Other spinal physical findings associated with thoracolumbar spinal injuries include spinal malalignment, spinous process gaps on palpation, spinal hematomas, and ecchymoses.
   - A thorough secondary survey should be performed. The patient should be logrolled and the entirety of the spine palpated. A rectal examination and assessment of perianal sensation can also be done with the patient logrolled.
   - Nonspine-related injuries are common and related to the mechanism of initial injury. These include fractured ribs, lung injury/contusion, pneumothorax, hemothorax, diaphragmatic rupture, hemopericardium, head injury, and injury to the great thoracic vessels.[1,2,3]
3. *Describe the three-column classification system (of Denis) for the thoracolumbar spine.*
   - Originally described by Francis Denis in 1983, the three-column model of the thoracolumbar spine allowed classification of thoracolumbar spine injuries based on CT scan.[4]
   - The anterior column was defined as the anterior longitudinal ligament (ALL) to the anterior two-thirds of the vertebral body, including the anterior two-thirds of the intervertebral disc.
   - The middle column as the posterior one-third of the vertebral body including the posterior longitudinal ligament (PLL) and the posterior one-third of the intervertebral disc
   - The posterior column as all structures posterior to the PLL (the pedicles, facet joints and articular processes, ligamentum flavum, and the neural arch/interconnecting ligaments).
   - See ▸ **Fig. 109.2** for an illustration.
4. *Name the six ligaments of the thoracolumbar spine and define which movements they limit.*
   - The six ligaments of the thoracolumbar spine include:
     - The ALL (limits extension)
     - The PLL (limits flexion)
     - The ligamentum flavum (limits flexion)
     - The supraspinous ligament (limits flexion)
     - The interspinous ligament (limits flexion)
     - The intertransverse ligament (limits lateral flexion)
5. *Describe the five relevant forces involved in structural damage to the thoracolumbar spine.*
   - Forces are:
     - Axial compression
     - Flexion/distraction
     - Hyperextension
     - Rotation
     - Shear[1]
   - Axial loading can result in compression fractures and burst-type fractures due to end plate failure and resultant compressive forces.
   - Flexion/distraction forces can result in pure osseous lesions, pure ligamentous or disc lesions, or mixed lesions.
   - Hyperextension can result in anterior discoligamentous injury with posterior element fractures.
   - Rotational injuries combine compressive loads and flexion/distraction mechanisms with the addition of rotational forces. Anterior and posterior element disruption can occur if rotational forces are significant enough to cause ligamentous and facet capsule integrity failure.
   - Shear injuries result in ligamentous disruption and frequently are associated with spinal cord injury caused by anterior spondylolisthesis.
6. *What are the advantages of early surgical stabilization compared to nonoperative management?*
   - The absolute indication for immediate surgical decompression is progressive neurological decline in the presence of neural element compression.
   - Early fixation of the unstable spine should be a priority following stabilization of potential life-threatening injuries.
   - Early intervention with surgical fixation allows for more rapid mobilization. This is the likely explanation for the significantly decreased length of ventilation and intensive care unit (ICU) care, overall hospitalization, as well as complications (including deep vein thrombosis [DVT] and infection) in operated patients.[5,6]
   - Other advantages of early surgical stabilization include[1]:
     - Earlier pain relief
     - Earlier return to work
     - Improved ability and ease in treating other injuries in polytrauma patients
   - Early surgery may independently improve outcomes after thoracolumbar spinal cord injury but this concept remains controversial. Early surgery may be difficult to achieve due to the presence of other injuries.[7]

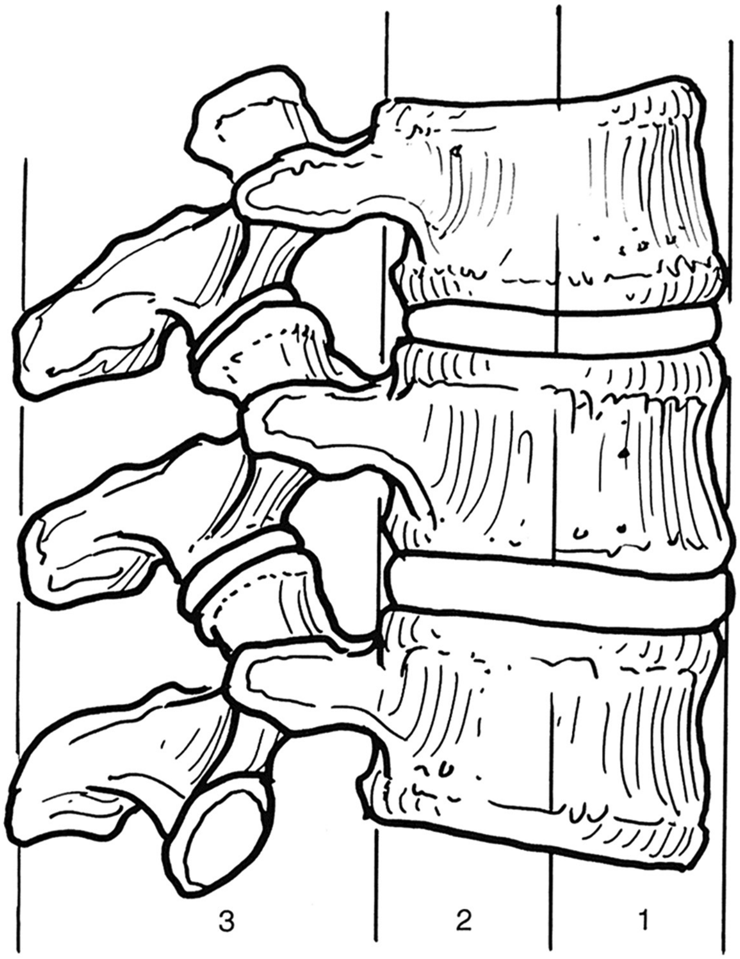

**Fig. 109.2** Schematic illustration of the three columns of Dennis. (Reproduced with permission from Dewald C. Spinal Deformities: A Comprehensive Text. New York: Thieme; 2003, p. 410)

## Answers (*continued*)

- However, there is no current class 1 evidence that suggests that patients without neurological deficit fare better with prophylactic early operative stabilization or maintain any long-term advantage.[8]

7. *Describe the major nonoperative and operative approaches to treat traumatic disc disruption and ligamentous injury.*
   - Nonoperative treatment of traumatic thoracolumbar disc disruption and ligamentous injury includes functional bracing, repositioning with cast immobilization, and physical therapy.
   - Operative approaches include posterior or anterior segmental reduction and stabilization, with or without decompression.
   - No definitive guidelines exist with regards to the approach or timing of surgical intervention including decompression in patients with acute spinal cord injury, though some data suggests that patients with acute spinal cord injury undergoing decompression surgery within 24 hours demonstrate greater improvement on the ASIA scale than those decompressed after 24 hours following injury.[9]
8. *What anatomical features of the thoracolumbar junction make it susceptible to injury?*
   - Factors to consider are the change from thoracic kyphosis to lumbar lordosis (occurring at T11–T12). The thoracic kyphosis will be reasonably rigid, whereas the lumbar lordosis is naturally more mobile.
   - While the first 10 ribs contribute to the relative stability of the thoracic column, the 11th and 12th, so-called free floating ribs, are not attached to the sternum and thus provide less stability at the thoracolumbar junction.
   - Facet joint anatomy is also relevant, as there is a transition from the more coronal plane of the facets in the thoracic spine to the more sagittal alignment in the lumbar spine.[1]

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# Case 110 Lumbar Burst Fracture

*Ahmed Alzahrani and Khalid Almusrea*

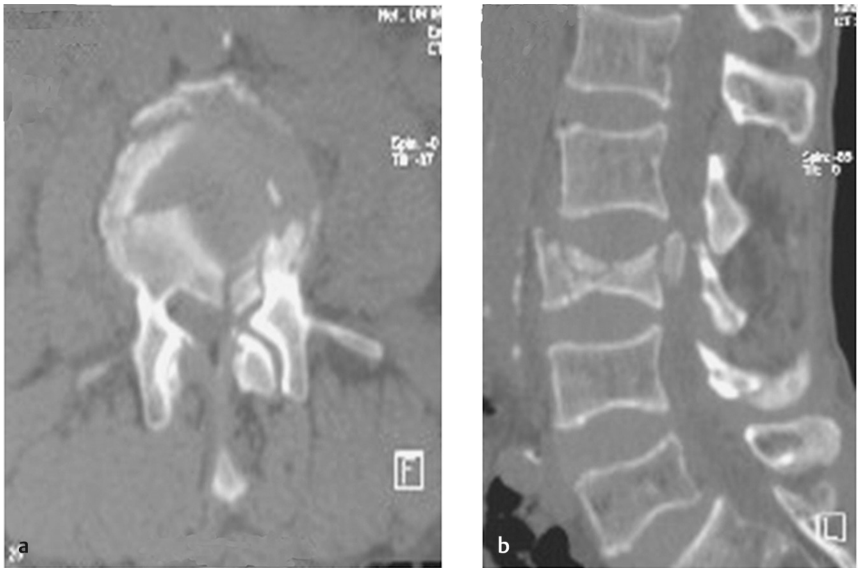

**Fig. 110.1** (**a**) CT scan of the lumbar spine with axial section through L1 and (**b**) sagittal reconstructed image.

## Clinical Presentation

- A 32-year-old woman presents after a fall from the ladder, with the complaint of severe low back pain and left foot pain.
- On examination, she is neurologically intact.
- She has an unremarkable past medical history.
- A CT scan of the lumbar spine was done (▶**Fig. 110.1**).

## Questions

1. Describe the CT images.
2. What is your initial management?

   MRI is obtained and pertinent images are shown in ▶**Fig. 110.2**.
3. What are the findings of the MRI of the lumbosacral spine?
4. What is TLICS scoring system?
5. What is your management now?
6. What are possible complications of nonoperative management in this case?
7. Describe indications for surgery in burst fractures.
8. What are the surgical options in this case?
9. What surgical approach will you choose and why?
10. What are the potential complications of a posterior surgical approach?
11. What are the outcomes of this approach?

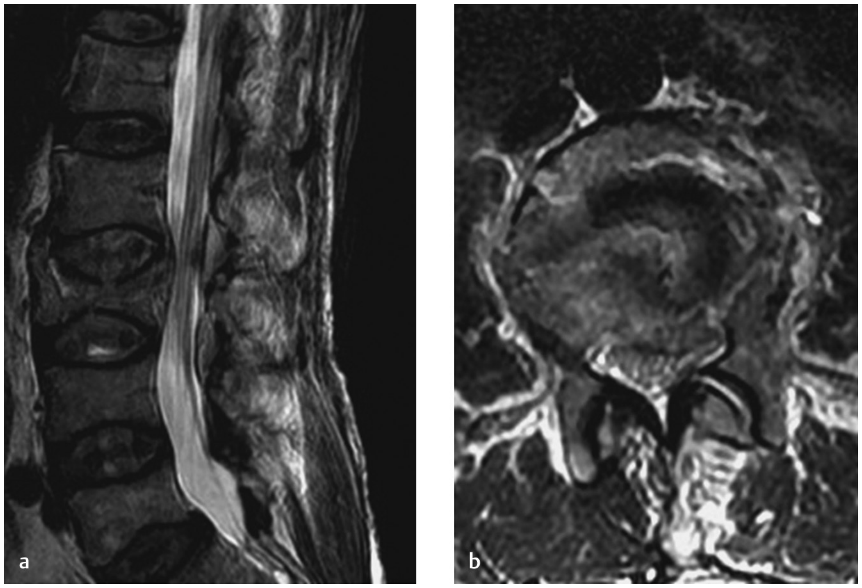

**Fig. 110.2** MRI scan of the lumbar spine with **(a)** T2-weighted sagittal section and (**b**) axial section images through L1.

## Answers

1. *Describe the CT images.*
   - There is a type A Denis burst fracture of the L1 vertebra.
   - Loss of height of ~50% and kyphosis of 15 degrees are evident.
   - There is canal compromise of ~30%.
2. *What is your initial management?*
   - Ensure the initial trauma work-up has been completed (airway, breathing, circulation [ABCs], etc.).
   - Complete spinal precautions (logroll only).
   - Obtain radiographs of the rest of the spine and further CT scans if other fractures are seen.
   - Place the patient on adequate pain management.
   - Obtain basic laboratory panel including complete blood count (CBC), electrolytes, coagulation profile.
   - Obtain an MRI of lumbosacral spine to assess any neural element and ligamentous involvement.
   - You may also elect to order an external orthosis, preferably a Jewett or thoracolumbar spinal orthosis brace in this case.
3. *What are the findings of the MRI of the lumbosacral spine?*
   - The MRI confirms the mild compromise of the thecal sac.
   - There is no intramedullary hyperintense signal (see **Case 107** for further details).
4. *What is TLICS scoring system?*
   - TLICS stands for Thoracolumbar Injury Classification and Severity Score.[1]
   - The classification system is based on three major categories: the morphology of the injury; the integrity of the posterior ligamentous complex; and the neurologic status of the patient.
5. *What is your management now?*
   - Treatment options include:
     - Conservative treatment with bracing, physical therapy, and pain control.[2]
     - Surgical fixation via either posterior minimal invasive or open approach.[3,4]
   - With the absence of significant canal stenosis and root compression, surgical intervention with minimal invasive approach may be a better option.
6. *What are possible complications of nonoperative management in this case?*
   - Persistent and worsening instability (mechanical, neurologic)
   - Increased kyphosis leading to neurologic deficit or intractable pain[5]
   - Further collapse of the affected vertebra with cauda equina syndrome
7. *Describe indications for surgery in burst fractures.*
   - Features of instability that lead to the requirement for surgical fixation include the following[5]:
     - Neurologic deficits
     - Canal compromise of ≥ 50%
     - Height loss of ≥ 40%
     - Kyphosis of ≥ 30 degrees
8. *What are the surgical options in this case?*
   - Minimal invasive spinal fixation and fusion from T12–L2
   - Open instrumented fusion T11–L2. An anterior, posterior or combined approach may be considered in this case. If a posterior approach alone is considered, then spanning the construct at least 2 levels above and 2 levels below the fracture should be considered, i.e., pedicle screw fixation from at least T11 to L3 should be performed as this fracture is by a junctional level.
9. *What surgical approach will you choose and why?*
   - Posterior minimal invasive fixation and fusion is preferred, because open posterior approach will need more muscle dissection for exposure despite

## Answers (*continued*)

no major difference in outcome or neurologic deficits that needs decompression of neurological elements.[4,6] (This is our personal preference; however, the choice of the approach is debatable and remains controversial.)

10. *What are the potential complications of a posterior surgical approach?*
    - In a series of 76 patients with thoracolumbar fractures, the complication rate was ~3.6%.[7]
    - Complications include the following[7–9]:
      - Pain
      - Neurologic deficit
      - Postoperative infection
      - Progression of the kyphosis
      - Nonunion or lack of fusion
      - Rupture or loosening of the implants
      - Loss of correction and required placement of an anterior support graft
      - Vertebral arch fracture
      - Epidural hematoma
      - Inadequate decompression
      - Cerebrospinal fluid leak
11. *What are the outcomes of this approach?*
    - Results are varied and depend on the studied parameters and institution.
    - In one study, no significant differences have been shown between the two approaches in terms of the functional and the radiological outcome 1 year after trauma when a posterior approach is used alone.[7]
    - The neurologic recovery from burst fractures is not predicted by the amount of initial canal encroachment or kyphotic deformity.[10]
    - In burst fractures without a neurologic deficit, there is no superiority of conservative therapy over operative therapy.[10] One prospective trial on 80 patients without neurologic deficits showed that posterior fixation provides partial kyphosis correction and earlier pain relief, but the functional outcome at 2 years is similar to a conservatively treated group.[11]
    - When there is significant neurologic involvement, operative management is advised. However, there is no obvious superiority of one approach over the other.[10]
    - In a series of 28 patients treated via posterior approach, there was 82% neurologic improvement.[9]
    - A retrospective review of 46 patients with encroachment of the spinal canal greater than 50% treated surgically showed that there is no significant difference in clinical outcome between those treated with a combined approach (anterior and posterior) versus those treated with a posterior approach alone. Furthermore, neurologic deficits improved by at least one Frankel grade in both cases for the most part.[12]

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# Case 111 Lumbar Chance Fracture

*Joaquin Hidalgo, Jared Marks, and Jorge E. Alvernia*

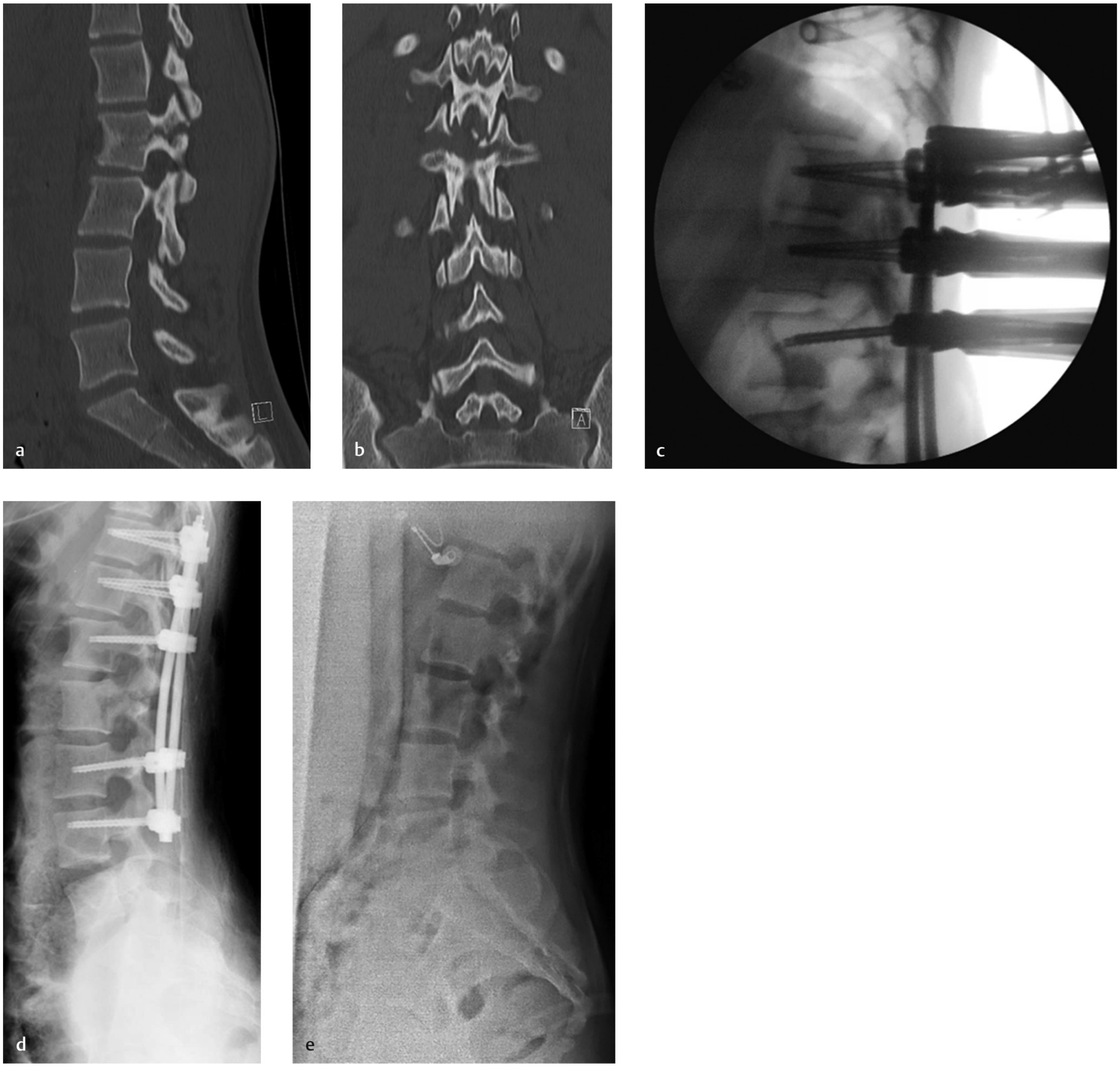


**Fig. 111.1** (**a**) Sagittal and (**b**) coronal reconstruction CT of the lumbar spine. (**c**) Intraoperative X-ray image during percutaneous minimally invasive pedicle screw placement. (**d**) Immediate postop X-ray image. (**e**) Five months after initial surgery, hardware was removed; the fracture line appears radiographically healed.

## Clinical Presentation

- An 18-year-old female restrained front passenger of a high-speed motor vehicle collision (MVC) presents to the emergency room. She complains of mild lumbar back and abdominal pain.
- On examination, she is neurologically intact. She has tenderness to palpation in the upper lumbar area. Radiological findings are shown in ▶**Fig. 111.1**a, b.

## Questions

1. Describe the images presented in ▶**Fig. 111.1**a, b.
2. What is a Chance fracture? What is the most commonly associated setting resulting in this injury?
3. What is the mechanism of this spine injury?
4. Where is the instantaneous axis of rotation located for the production of a Chance fracture?
5. What is the most common level of the spine associated with Chance fractures?
6. What is the most commonly affected age group and what is the most common neurological presentation of this fracture?
7. What are the morphological variants of this fracture? How can they be classified according to morphology and stability?
8. What other injuries can be associated, leading to what further work-up?
9. What are the treatment options for this fracture?
10. What are the potential complications that could arise in long-term follow-up?

## Answers

1. *Describe the images presented in* ▶**Fig. 111.1***a, b.*
   - Plain CT of the lumbar spine shows:
     - Horizontal fracture involving the vertebral body, pedicle, and pars interarticularis of L2
     - Coronal view demonstrates complete splitting of the posterior bony elements of the L2 vertebra.
2. *What is a Chance fracture? What is the most commonly associated setting resulting in this injury?*
   - Described by George Quentin Chance in 1948 as: “Horizontal splitting of the spine and neural arch and vertebral body with no vertebral body wedging or apophyseal joint dislocation.”[1]
   - After the first description of this fracture, its association with the use of lap seatbelts by passengers in motor vehicle accidents was made. As a consequence, the terms “seatbelt fracture” and “Chance fracture” are sometimes used interchangeably for spinal fractures resulting from similar mechanisms as the classic Chance fracture, but with different morphologies.[2]
3. *What is the mechanism of this spine injury?*
   - The mechanism of injury of Chance fractures is thought to be *flexion-distraction* of the spine,[2,3] usually due to sudden deceleration of the body with the pelvis or anterior abdominal wall restrained, resulting in forward hyperflexion of the torso around the restraining object. This generates tensional disruption (distraction) of the elements of the spine.[4]
4. *Where is the instantaneous axis of rotation located for the production of a Chance fracture?*
   - The physiologic flexion axis of rotation of the thoracolumbar spine is at a point located near the center of the intervertebral disc space.[2,3]
   - During the hyperflexion phase, the instantaneous axis of rotation is moved to a point anterior to the vertebral body, where the restraining object is in contact with the pelvis or abdominal wall and functions as a fulcrum.[2] The classically reported object serving as fulcrum is a lap seat belt; however, it can occur in any situation that emulates the same mechanism (for example, fall from a height with the torso forced into flexion over a fence).[2]
5. *What is the most common level of the spine associated with Chance fractures?*
   - Although this mechanism of injury can occur at any level of the spine, it is more commonly seen in the thoracolumbar spine (T11–L2) due to the high mobility of this segment, as compared to the thoracic spine where the rib cage limits the mobility.[4,5]
6. *What is the most commonly affected age group and what is the most common neurological presentation of this fracture?*
   - The incidence of this type of fracture is higher in the pediatric population. This is due to the child’s head–body weight ratio and the resultant center of gravity is higher, leading to a greater lever arm during the hyperflexion phase and greater tensile forces within the spine.
   - Around 75% of patients with a flexion-distraction spine injury will present with no neurological deficits at all (ASIA score E). Ten percent will present with a complete spinal cord injury (ASIA score A), and the rest will present as various degrees of incomplete spinal cord injury (ASIA scores B, C, or D).[5]

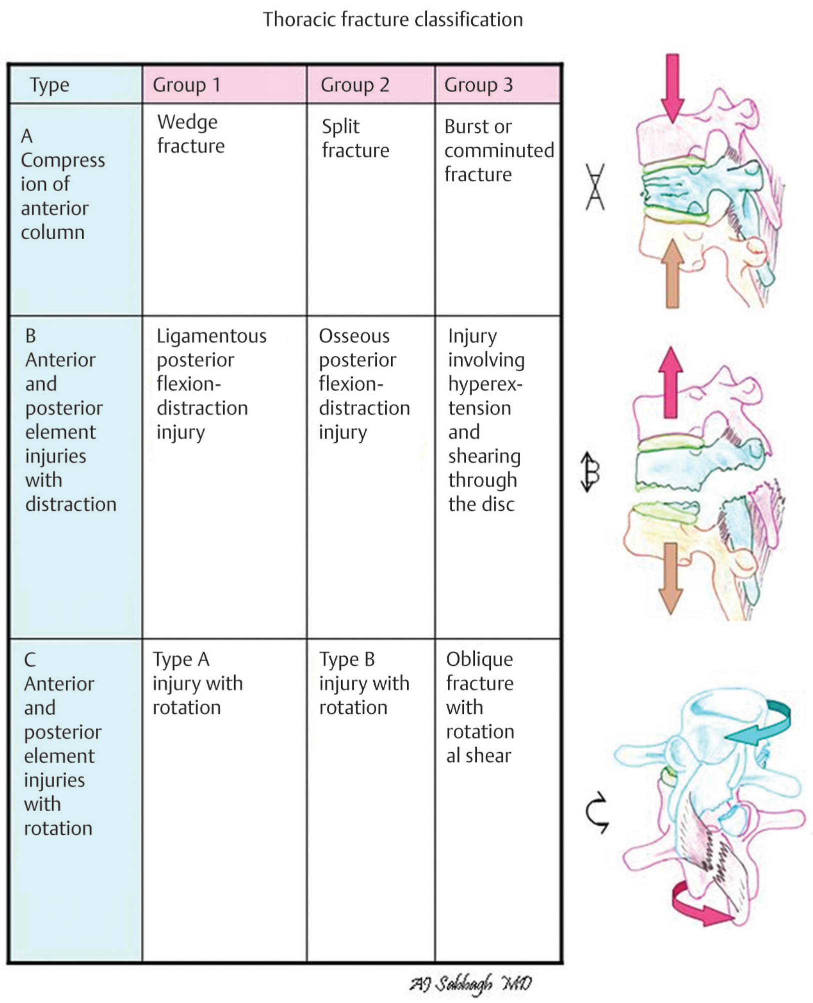
## Answers (*continued*)

7. *What are the morphological variants of this fracture? How can they be classified according to morphology and stability?*
    - The morphology of thoracolumbar fractures resulting from flexion-distraction mechanisms can be quite variable. The horizontal “splitting of bone only” in the classic Chance fracture can often be accompanied by vertebral body wedge deformity or burst fractures. Furthermore, no bony involvement with tensional failure of discoligamentous elements only is also possible, but less common.[2,5]
    - Flexion-distraction spine injuries including the Chance fracture are described as *unstable* injuries with failure of both the posterior and middle columns due to tensile forces.[6] There is no consensus in the literature as to whether the anterior column also fails in tension or compression (Chance fractures associated with wedge deformity or comminution of the vertebral body).[4]
    - There are numerous classifications for traumatic spine injuries. One of the most comprehensive and more recently developed is the AOSpine Thoracolumbar Injury Classification System (TLICS; ▶**Table 111.1**). In this classification, flexion-distraction injuries are graded as type B with three subtypes, illustrated in ▶**Fig. 111.2**.[7]
    - This patient could be classified morphologically as posterior transosseous tension band disruption (type B.2. of the AOSpine TLICS).
8. *What other injuries can be associated, leading to what further work-up?*
    - There is a high incidence of associated intra-abdominal injuries, including intestinal perforation, vascular injury, and visceral laceration. The “seat belt sign” can herald these underlying injuries[2,5,8,9] and should be looked for on examination.
    - Rigorous examination and CT scan of the abdomen should be completed, and trauma surgery consultation should be considered.
9. *What are the treatment options for this fracture?* Options are as follows:
    - *Nonoperative treatment:* immobilization in hyperextension with orthosis. Preferred for neurologically intact patients without ligamentous involvement and minimal kyphotic deformity (< 17°). Requires patient suitability for bracing, adherence to treatment, and close radiographic follow-up.[10]
    - *Surgical stabilization:* Pedicle screw instrumentation, open or percutaneous, is the preferred surgical approach. It is indicated in the setting of ligamentous disruption, > 17 degree kyphotic deformity, neurological compromise, and failure or noncompliance with external immobilization. Either a short or long segment construct can be used.[11,12]
    - *In the presence of thecal sac compression,* decompression through posterior, transpedicular, anterior or combined approach should be considered, since reduction of the fracture would result in increased compression.
10. *What are the potential complications that could arise in long-term follow-up?*
    - Wound infection can be more prevalent in patients with associated bowel perforation.[5] Posttraumatic kyphosis is almost exclusively seen in patients treated nonoperatively, and > 5 degree increase in angulation should be treated surgically. Important instrumentation-related complications include pain and prominence (18%), and implant breakage (< 1%). The hardware can be removed in these patients if the fracture tract has healed radiographically.[5,11,12]

**Table 111.1** AOSpine Thoracolumbar Fracture Classification

| | |
|---|---|
| A. Compression fractures of the vertebral body | A.1. Wedge fracture |
| | A.2. Split vertebral body fracture |
| | A.3. Incomplete burst fracture |
| | A.4. Complete burst fracture |
| B. Tension band injuries | B.1. Posterior transosseous disruption |
| | B.2. Posterior ligament disruption |
| | B3. Anterior ligament disruption |
| C. Translation injuries | |

Adapted from Vaccaro et al 2013.[7]



Thoracic fracture classification

| Type | Group 1 | Group 2 | Group 3 |
|---|---|---|---|
| A<br>Compression of anterior column | Wedge fracture | Split fracture | Burst or comminuted fracture |
| B<br>Anterior and posterior element injuries with distraction | Ligamentous posterior flexion-distraction injury | Osseous posterior flexion-distraction injury | Injury involving hyperextension and shearing through the disc |
| C<br>Anterior and posterior element injuries with rotation | Type A injury with rotation | Type B injury with rotation | Oblique fracture with rotational shear |

**Fig. 111.2** Thoracolumbar fracture classification. The subtypes of tension band injuries: B1, transosseous disruption; B2, posterior tension band injury; B3, anterior tension band injury. (Reproduced from Nader et al. Neurosurgery: Tricks of the Trade. New York: Thieme; 2010)

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# Case 112 Gunshot Injuries to the Spine

*Luke G. F. Smith and H. Francis Farhadi*

## Clinical Presentation

- A 28-year-old male presents as a level I trauma after an altercation that led to him being shot in the posterior neck twice.
- On examination, he has intact deltoid function bilaterally but has 3/5 bilateral elbow flexion, and no further movement in the rest of the muscle groups in both upper and lower extremities. Intact sensation is noted over the lateral upper arms while no sensation is evident over the rest of the upper extremities and over the chest. No sensation is noted to Foley catheter manipulation and there is no rectal tone.

## Questions

1. What are the ASIA grade and level?
2. Describe the CT imaging findings.
3. What demographic is most afflicted by gunshot injuries to the spine? What segment of the spine is most commonly injured?
4. What are the two acute types of damages caused by projectiles to the spinal cord? Describe the mechanisms of both.
5. List the critical early evaluation and management measures following a gunshot injury to the spine.
6. Describe the algorithm for tetanus prophylactic vaccination.
7. For how long should antibiotics be given following gunshot injury to the spine?
8. Are steroids recommended in the treatment of penetrating injury to the spinal cord?
9. What are the indications for surgical intervention following gunshot injury to the spine?
10. What is the role of MRI following gunshot injury to the spine?
11. What is the most important predictor of lead toxicity with a retained bullet fragment? By what method does lead leech from the retained fragment?
12. Is a gunshot injury or nonviolent spinal cord injury more likely to lead to complete injury?

## Answers

1. *What are the ASIA grade and level?*
   - ASIA A at a C5 level
2. *Describe the CT imaging findings.*
   - CT scan of the cervical spine shows retained bullet fragments evident at the C6 and C7 levels (▶**Fig. 112.1**a).
   - Destruction of the left C7 lateral mass and left laminar fracture is seen (▶**Fig. 112.1**b).
3. *What demographic is most afflicted by gunshot injuries to the spine? What segment of the spine is most commonly injured?*
   - Most victims are males, 78 to 91% in some studies, regardless of civilian or military designation, with the highest incidence in the third decade of life.
   - The thoracic spine is affected most commonly followed by the cervical and lumbar spine.
4. *What are the two acute types of damages caused by projectiles to the spinal cord? Describe the mechanisms of both.*
   - The two general types of damage include direct mechanical injury via the projectile and indirect injury through shock waves or cavitation from high-energy projectiles.
   - Projectiles must have a velocity greater than 2,000 to 3,000 feet/second in order to cause indirect injury, which includes projectiles from military assault rifles and other high-powered guns.
   - There have been reported cases of spinal cord injury through indirect mechanisms without breach of the spinal canal.
5. *List the critical early evaluation and management measures following a gunshot injury to the spine.*
   - Maintenance of ABCs (airway, breathing, and circulation) and assessment of potential life-threatening injuries are the primary initial goals.
   - Tetanus prophylaxis must be considered in all instances.

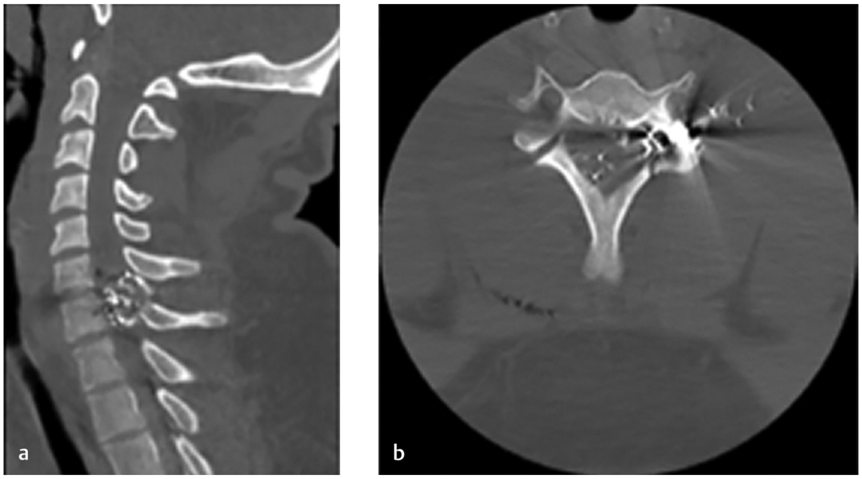

**Fig. 112.1** Sagittal (**a**) and axial (**b**) CT views of the cervical spine show intracanal and intramedullary bullet fragments.

## Answers (*continued*)

- Broad-spectrum antibiotics should be initiated as soon as possible.
- Entrance and exit wounds should be examined thoroughly. The bullet entry site should be identified and treated with debridement of any devitalized skin and superficial soft tissues.
- Spinal instability should be assessed with lateral flexion-extension cervical radiographs in cooperative and neurologically intact patients. Severe comminution of anterior and/or posterior elements with evidence of segmental abnormal angulation or translation is indicative of instability.
- New-onset or progressive neurological deficit associated with an intramedullary bullet, bone fragment, or expanding epidural hematoma is an indication for urgent decompression. Lead intoxication from the bullet or contact of a copper jacketed missile with the neural elements are rare indications for surgical intervention.
- A role for prophylactic irrigation and debridement, particularly for associated viscus perforation, has been evaluated but remains to be defined.
- A lumbar subarachnoid drain should be placed if a cerebrospinal fluid (CSF) leak appears to be present.

6. *Describe the algorithm for tetanus prophylactic vaccination.*
   - All individuals with an unknown vaccination history or who have received < three doses of the tetanus vaccine should receive tetanus immunoglobulin.
   - If three doses of the vaccine were given < 5 years ago, no vaccine or immunoglobulin is needed.
   - If the three doses were given and it has been more than 5 years since the last dose, then the Tdap tetanus toxoid vaccine is recommended.
7. *For how long should antibiotics be given following gunshot injury to the spine?*
   - There are no consistent recommendations for antibiotics in the setting of gunshot wounds to the spine.
   - For gunshot wounds not complicated by viscus perforation, it is generally recommended to maintain 48 to 72 hours of prophylaxis.
   - If the injury involves an abdominal viscus perforation, there is increased risk of meningitis and sepsis. A minimum of 7 days broad-spectrum antibiotics is associated with lower infection rates.
   - The presence of bullet fragments has no association with increased infection risk. There is a higher risk of complications from attempted removal of fragments as compared to potential infection control benefits in the setting of perforated abdominal organs.
8. *Are steroids recommended in the treatment of penetrating injury to the spinal cord?*
   - Steroid administration is not recommended following a penetrating spinal cord injury. While no neurological improvement has been demonstrated, there is increased risk of wound breakdown, infection, and sepsis.
9. *What are the indications for surgical intervention following gunshot injury to the spine?*
   - The four main indications of surgery include progressive loss of neurological function, spinal instability, repair of persistent CSF leak, and bullet removal.
   - Neurological decline is a reason to emergently decompress the afflicted spine. Delayed neurologic decline has been reported with bullet migration within the canal.
10. *What is the role of MRI following gunshot injury to the spine?*
    - The presence of retained fragments is a relative contraindication to MRI. Although most bullets made in the United States are nonferromagnetic, some bullet jackets contain metals that could interact with MRI scanners.
    - While not routinely obtained in the acute setting of gunshot injury to the spine, MRI may help determine if a fragment is causing progressive decline

## Answers (*continued*)

and if surgical intervention is warranted. A discussion about risks of undergoing MRI should be undertaken with patients in this setting.

11. *What is the most important predictor of lead toxicity with a retained bullet fragment? By what method does lead leech from the retained fragment?*
    - Bullet location is the most important indicator of potential systemic lead poisoning.
    - While more common in synovial joints where the lead is dissolved by synovial fluid, lead poisoning has been reported with bullet fragments lodged in the intervertebral disc.
12. *Is a gunshot injury or nonviolent spinal cord injury more likely to lead to complete injury?*
    - Spinal cord injury caused by gunshot is more likely to cause a complete injury and cause paraplegia compared to nonviolent injury.

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# Case 113 Spinal Cord Injury without Radiologic Abnormality (SCIWORA)

*Jared Fridley, Andrew Jea, and Ibrahim Omeis*

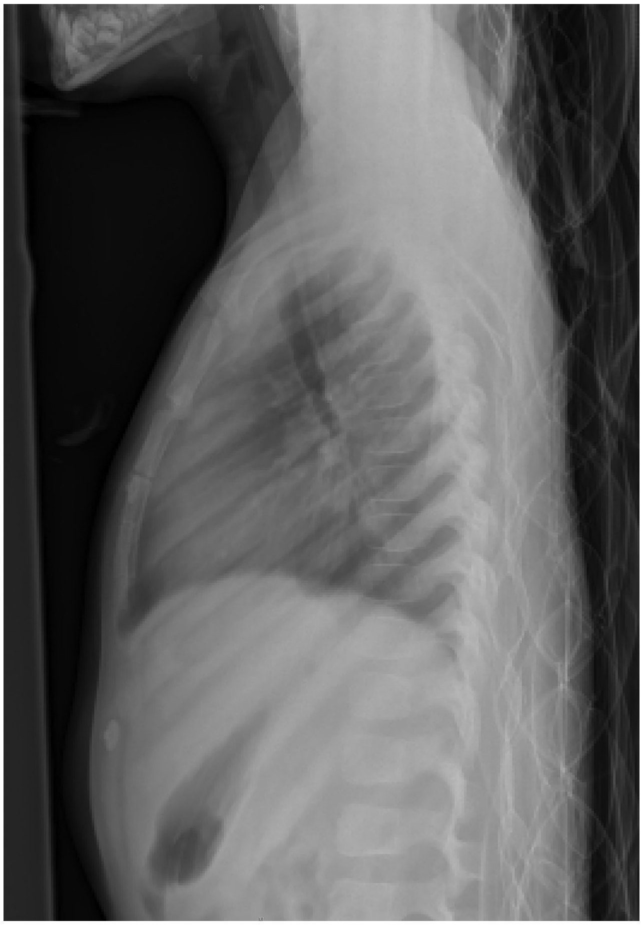

**Fig. 113.1** Lateral radiograph of the thoracic spine.

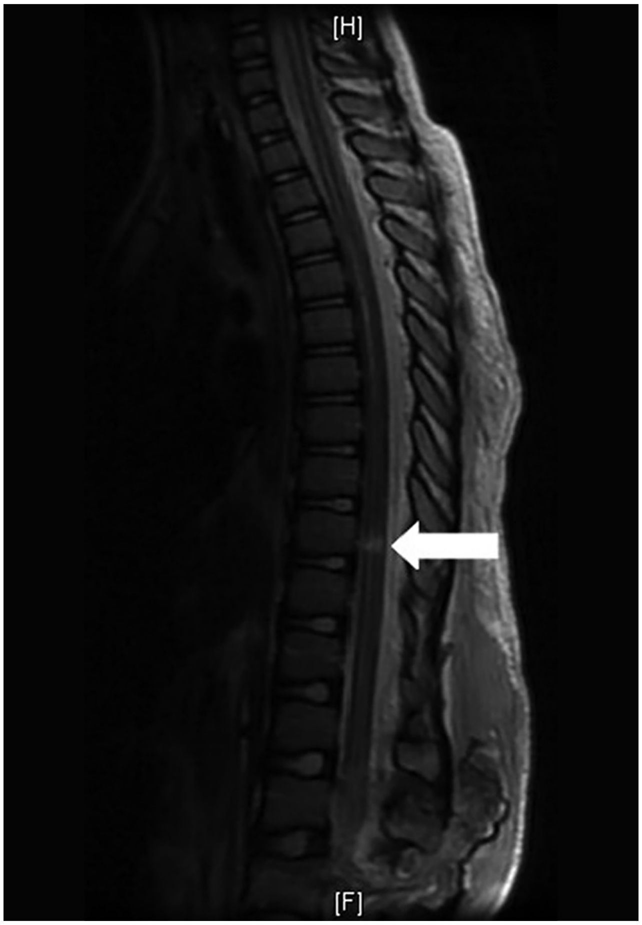

**Fig. 113.2** MRI thoracic spine T2-weighted image, midsagittal view.

## Clinical Presentation

- A 6-year-old girl presents to the emergency room (ER) after a high-speed motor vehicle accident, awake and alert, but unable to move her legs.
- Neurologic examination reveals complete paraplegia of the lower extremities and T10 sensory level.
- Supine thoracic spine radiographs were obtained in the ER (▶**Fig. 113.1**).

## Questions

1. Interpret the lateral thoracic spine radiograph (► **Fig. 113.1**).
2. What is your differential diagnosis?
3. An MRI scan of the spine without contrast was obtained. Interpret the image (► **Fig. 113.2**).
4. What is spinal cord injury without radiologic abnormality (SCIWORA)?
5. Which group(s) of patients are most prone to SCIWORA and why?
6. What is the diagnostic work-up of pediatric patients with evidence of spinal cord injury?
7. What is the management for SCIWORA?
8. What is the prognosis of patients with SCIWORA?
9. What are some of the delayed complications possible in a child with SCIWORA?
10. Discuss the embryology of the developing pediatric spine and how this can facilitate the development of SCIWORA?

## Answers

1. *Interpret the lateral thoracic spine radiograph (►* ***Fig. 113.1****).*
   - These radiographs demonstrate no gross fracture or malalignment of the thoracic spine.
2. *What is your differential diagnosis?*
   - Traumatic myelopathy due to hematoma, fracture, or disc herniation.
   - SCIWORA. Term used to describe a patient with evidence of spinal cord injury on examination, but no acute abnormalities on spine radiographs or CT scan.
   - Acute demyelinating encephalomyelitis
   - Acute disseminated encephalomyelitis
   - Transverse myelitis
   - Vertebral artery traumatic/nontraumatic thrombosis
3. *An MRI scan of the spine without contrast was obtained. Interpret the image (►* ***Fig. 113.2****).*
   - This sagittal T2-weighted MRI sequence shows a focal hyperintensity at the T10 level, suggestive of spinal cord injury. There is no evidence of hematoma, disc herniation, or fracture.
4. *What is spinal cord injury without radiologic abnormality (SCIWORA)?*
   - SCIWORA refers to a patient with evidence of trauma-related myelopathy, without evidence of spinal fracture, dislocation, or ligamentous injury.[1] Occlusion of the vertebral arteries may cause injury to the spinal cord. The establishment of the cervical vertebrae back to their normal alignment provides removal of the "occlusion" and flow restoration to the vertebral arteries.
5. *Which group(s) of patients are most prone to SCIWORA and why?*
   - SCIWORA usually occurs in a bimodal distribution in the pediatric population between infancy and adolescence, and in older adults.
   - While spinal cord injury in the pediatric population is rare, the immature skeletal structures of children make them uniquely prone to SCIWORA, particularly in the cervical spine. This is due to the elasticity of the spinal ligaments, an underdeveloped paraspinal musculature, relatively large head size, and the horizontal orientation of facet joints.[1]
   - SCIWORA has been reported to occur in nearly two-thirds of children who present with closed spine injury, usually due to motor vehicle-related trauma or falls from height.[2,3]
   - Older adults with SCIWORA usually present with central cord syndrome, often secondary to hyperextension injuries in the context of significant spondylotic stenosis.
6. *What is the diagnostic work-up of pediatric patients with evidence of spinal cord injury?*
   - Following initial resuscitation (ATLS airway, breathing, circulation [ABCs]), and spinal immobilization, a thorough history, if possible, is obtained from family or witnesses at the scene of the injury to help ascertain the mechanism of trauma.
   - In general, cervical spinal radiographs are obtained for children who are unable to communicate; have a neurological deficit, neck pain, a painful distracting injury; are intoxicated; or sustain a fall from height, motor vehicle accident, or nonaccidental trauma as the mechanism of injury.[4] Additional radiographs of the thoracic and/or lumbar spine are obtained, if indicated, based on examination.
   - If no spine fractures or evidence of overt instability are visualized on radiographs, then a CT scan of the spine region of interest is performed to look for occult fractures.
   - A spine MRI is obtained as soon as possible to rule out a traumatic disc herniation, intraspinal hematoma, or evidence of ligamentous injury. If an MRI is unable to be obtained, dynamic flexion-extension radiographs are performed, though usually in a delayed fashion due to muscle spasm.
7. *What is the management for SCIWORA?*
   - The primary goal of treatment is to prevent further spinal cord injury.[5] This includes immobilization of the affected segment, avoidance of exacerbating activities, and blood pressure support as needed to allow adequate spinal cord perfusion.[6]

## Answers (*continued*)

- Delayed presentation of SCIWORA can occur from 30 minutes to 4 days after injury, as well, cases of recurrent SCIWORA weeks after injury have been reported in the literature.[7] In both delayed and recurrent SCIWORA, occult instability coupled with exacerbation of injury due to activity and/or removal of an external orthotic is thought to have occurred.
- Administration of a short course of methylprednisolone for children within 8 hours of severe spinal cord injury is practiced at some centers,[7] though there is little published data in pediatric patients to support this practice.

8. *What is the prognosis of patients with SCIWORA?*
   - Twenty to 30% of patients with SCIWORA have a complete cord injury,[1] usually in the lower cervical or upper thoracic spine. Similar to adult patients, children with complete injury rarely improve, while those with mild to moderate spinal cord injury will often fully recover. Children under the age of 8 years generally have the worst prognosis.[8]
   - MRI findings within the spinal cord may be predictive of prognosis and a more useful biomarker for outcome than initial neurologic examination. Those with transection or major hemorrhage have a poor outcome with little chance of recovery, while those with minor hemorrhage or only T2 hyperintensity tend to have a good prognosis for recovery.[9] Even in patients with a severe injury on initial neurologic examination, those with only T2 hyperintensity in the cord at the level of injury tend to have a good recovery.
9. *What are some of the delayed complications possible in a child with SCIWORA?*
   - Children diagnosed with SCIWORA should be followed diligently to anticipate any potential complications which may include spinal deformity, syringomyelia, and another, but delayed SCIWORA, termed "second SCIWORA," which will usually present by 2 weeks post injury.[10]
10. *Discuss the embryology of the developing pediatric spine and how this can facilitate the development of SCIWORA?*
    - The key point to remember when interpreting cervical spine radiographs in the young pediatric population is the presence of synchondroses, joining the various ossification centers. A common pitfall is to interpret these normal synchondroses as fracture lines.
      - In considering C1, for instance, three ossification centers, representing each of the lateral masses and the anterior arch, will completely fuse by 7 years of age. The posterior arch, though, often retains a bifid appearance due to lack of fusion in the midline, and this can easily be interpreted erroneously as a fracture line.
      - In C2, a cartilaginous physis joins the odontoid to the body and can often be mistaken for a fracture up to the age of 5 to 7 years. The very tip of the dens will fuse via the ossification center, the ossiculum terminale, which manifests at 7 years of age, then fuses to the dens by 12 years of age.
      - The remainder of the cervical vertebrae all have similar formation patterns, with one ossification center for the body and another for the associated posterior arches and lateral masses. These will unite in the midline around 2 to 4 years of age. The neurocentral synchondrosis will close between 3 and 6 years of age.[10]
    - A thorough understanding of the neuroanatomy of the developing pediatric cervical spine is paramount to correctly interpreting imaging and formulating the safest, most appropriate course of treatment for the child.

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# Case 114 Acute Cervical Disc Herniation

*Juan Ortega-Barnett*

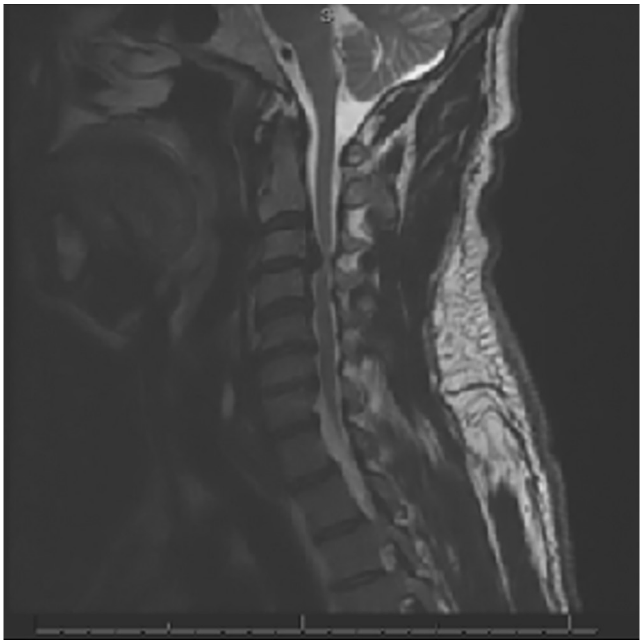

**Fig. 114.1** MRI T2-weighted image, sagittal view of the cervical spine.

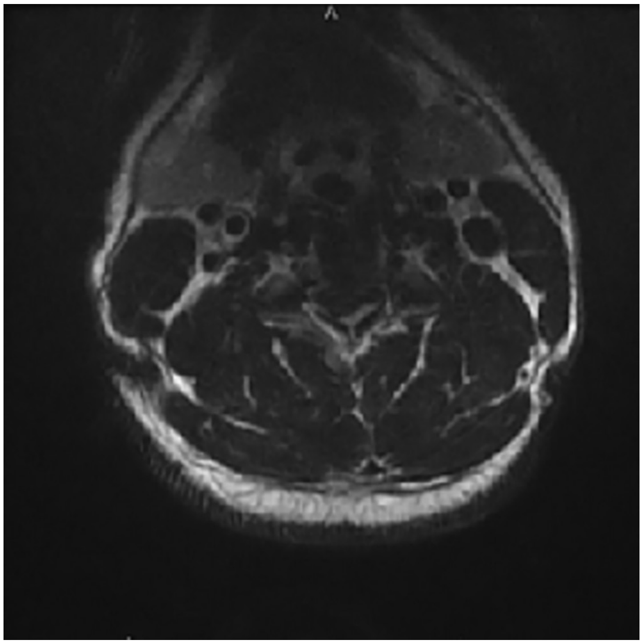

**Fig. 114.2** MRI T2-weighted image, axial view of the cervical spine.

## Clinical Presentation

- A 51-year-old Hispanic male reports that over the past several years he has had neck pain. During the previous couple of months, he noticed increased weakness and stiffness in the limbs on the right half of his body that has been getting progressively worse. He has noticed a right foot drop. He reports progressively declining dexterity in his right hand and decreased sensation in all of the fingers of that hand. He reports that if he sits for a few minutes, he has incredible stiffness and difficulty with mobility, requiring him to stretch and warm up before he can move. He says that his right arm tightens uncontrollably and twists when he stands. When he walks, he occasionally observes that he drags his right foot behind him.
- His neck pain became severe during the past 2 weeks and he had worsening of his previously described symptoms. He denies any bowel or bladder incontinence. He denies any stroke or history of seizure. He has an extensive history of intravenous (IV) heroin use and is hepatitis C positive.
- On exam, the patient has increased muscle tone with right hand grasp weakness and forearm flexion 4/5, with right lower extremity dorsiflexion weakness 3/5, and hyperreflexia.
- See ▸**Fig. 114.1** and ▸**Fig. 114.2** for imaging studies.

## Questions

1. Describe the MRI findings.
2. Describe the clinical signs and symptoms of C4 radiculopathy.
3. What is your differential diagnosis?
4. What is your management?
5. Describe the risks and potential complications of an anterior cervical discectomy and fusion (ACDF).
6. The patient develops a postoperative hematoma and has difficulty breathing while in the recovery room. Describe your management at this point.

## Answers

1. *Describe the MRI findings.*
   - This is a T2-weighted MRI of the cervical spine with sagittal and axial views. At C3–C4, there is advanced degenerative changes including superior migration of a central disc extrusion with uncovertebral joint hypertrophy, and facet joint degeneration results in severe spinal canal stenosis and mild bilateral neural foraminal narrowing at this level.
   - Increased T2 signal along the spinal cord adjacent to the C3–C4 level likely represents myelomalacia/compressive myelopathy secondary to severe compression at this level. At levels C4–C5 through C6–C7, there is degenerative disc disease with osteophyte complexes and mild canal narrowing.
2. *Describe the clinical signs and symptoms of C4 radiculopathy*.
   - A C4 radiculopathy may produce nonradiating axial neck pain. Patients may also present with trapezius pain.[1]
   - More commonly, neck extension aggravates the pain more so than flexion but generally speaking most cervical spine disc herniations cause painful limitation of neck movement.
   - When the examiner exerts downward pressure on the vertex while tilting head toward symptomatic side, this may reproduce the pain (Spurling's sign). Some patients find relief when elevating the arm and holding or placing the hand behind the head or on the vertex (abduction relief sign). Occasionally, electrical shock-like symptom traveling down the spine may be present (L'hermitte's sign).
3. *What is your differential diagnosis?*
   - Acute cord compression presenting with myelopathy and symptoms of central cord syndrome or even Brown–Sequard syndrome is well described in traumatic cervical disc herniation but is less common in nontraumatic cervical disc herniation.[2–4]
   - Differential causes of neck pain should include cervical sprain, fracture of the cervical spine, cervical spondylosis including facet arthritis, disc herniation, occipital neuralgia, atlantoaxial subluxation, Chiari I malformation.
   - In cases where radiculopathy is present, consideration for possible shoulder pathology should be excluded. Usually diseases of the shoulder do not produce referred pain to the neck.
   - Some interscapular and shoulder pain may be referred from cholecystitis. Some cases of left C6 cervical radiculopathy may suggest acute myocardial infarction. Complex regional pain syndromes may benefit from stellate ganglion blocks.[5]
4. *What is your management?*
   - Most patients with nonsignificant chronic cervical spine disease and an acute cervical radiculopathy due to cervical disc herniation can improve without surgery.[6]
     – Medical management would include pain medication, nonsteroidal anti-inflammatory medication (NSAID's), and/or short-course tapering steroids.
     – However, recent studies do not support the use of high-dose steroids in cases of blunt spinal cord traumatic injuries.[7]
     – A cervical soft collar may be utilized temporarily. However, prolonged use of a collar leads to deconditioning of the neck musculature and tissue damage and should be avoided.
     – In general, studies that describe nonoperative therapy discuss the usage of multiple modalities of treatment. These include range-of-motion exercises, ergonomic neck instruction, neck relaxation, and superficial heat and manual cervical traction.
     – Alternative modalities include: epidural steroid injections, transcutaneous electrical nerve stimulation (TENS), cervical pillow, massage, acupuncture, and ultrasound.[1]
   - Surgery is indicated for those who fail to improve or those who have worsening neurological symptoms or present with myelopathy/central cord syndrome that is associated with acute cervical disc herniation.
   - There does exist controversy about surgical timing for acute cervical myelopathy and some cases have been reported with favorable outcome during their natural history course.
   - Surgical options include anterior approach with cervical discectomy with or without a prosthesis and fusion, and posterior decompression by laminectomy, laminoplasty, or laminotomy.
5. *Describe the risks and potential complications of an anterior cervical discectomy and fusion (ACDF).*
   - Complications that may be encountered when performing an ACDF include: vocal cord paresis from recurrent laryngeal nerve. Symptoms may include hoarseness, cough, aspiration, and dysphagia. Injury may occur from prolonged self-retaining retractors compressing the nerve against the trachea. This is more commonly observed when performing a right-sided approach.
   - Direct injuries may occur to the vertebral artery, jugular vein, and the carotid artery. Although usually the carotid may suffer occlusion, thrombosis, or laceration from retraction or excessive manipulation.
   - Perforation of the esophagus or trachea are minimized by careful dissection and retraction of the soft tissues. One should avoid keeping the drill running when exiting or entering the disc space work area.
   - If a dural tear is unsuccessfully repaired, this may lead to a cerebrospinal fluid (CSF) fistula.

## Answers (*continued*)

- Injury to the sympathetic plexus within the longus coli may lead to a Horner's syndrome.
- Thoracic duct injury will happen more often on a left-sided approach to the lower end of the cervical spine.
- Spinal cord injury may occur in patients with significant cervical canal stenosis and hyperextension during intubation. Care must also be taken when placing the bone graft which must fit within the limits of the adjacent vertebral body end plates. Excessive force applied when tapping the graft into place could cause spinal cord injury.
- Other complications include pseudoarthrosis which occurs more often in smokers. Graft extrusion and anterior angulation deformity are also known complications.
- Wound infections are less common than posterior cervical approaches but are reported to be between 1.2 and 3.2%.[8]
- Postoperative hematomas should be avoided by careful hemostasis and wound drain placement when indicated. When working at lower cervical levels, risk of pneumothorax and hemothorax exists.
- Adjacent degenerative disc disease may represent a sequela to altered biomechanics from surgery or a predisposition to cervical spondylosis. Many may be asymptomatic.[9]

6. *The patient develops a postoperative hematoma and has difficulty breathing while in the recovery room. Describe your management at this point.*
   - The priority is to establish and maintain patency of the airway. This is achieved by intubation by direct laryngoscopy, fiberoptic bronchoscopy, or by surgical means.
   - The patient is placed initially on supplemental oxygen delivery: 100% oxygen via nonrebreather facemask.
   - IV sedatives and pain killers are held at this time so as not to depress the respiratory drive.
   - If direct laryngoscopy is chosen, one should be aware that the anatomic landmarks and endotracheal tube passage can be challenging due to edema of the pharynx, epiglottis, and vocal cords. Repetition of failed intubation attempts should be avoided.
   - A delay in the establishment of an airway prolongs the period of hypoxia and can result in cerebral ischemia.
   - If a surgical airway is needed, a cricothyroidotomy represents a rapid method of obtaining an airway.
   - Next, the patient is taken back to the operating room (if feasible in a timely fashion, otherwise the next steps are immediately undertaken at the bedside); the incision is reopened and any hematoma should then be evacuated from the space between the carotid sheath and the midline viscera. A drain should be placed at this time. Closure is then achieved in a standard fashion.
   - Postoperatively, the patient should be transferred to the intensive care unit, intubated, and should remain intubated overnight until it is confirmed that the edema surrounding the trachea has subsided.[10–12]

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# Case 115 Anterior versus Posterior Approaches to the Cervical Spine

*Amgad S. Hanna and Kimberly Hamilton*

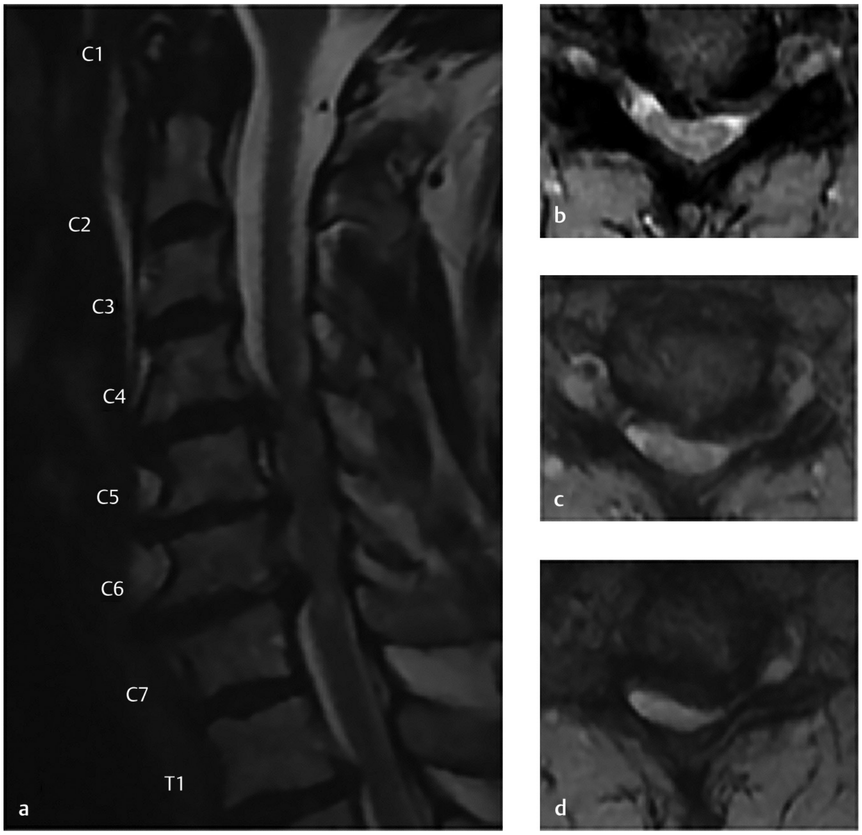

**Fig. 115.1** Preoperative imaging. (**a**) Cervical spinal T2-weighted MRI, midline sagittal image; (**b**) Cervical spinal T2-weighted MRI, C4–C5 axial image; (**c**) Cervical spinal T2-weighted MRI, C5–C6 axial image; (**d**) Cervical spinal T2-weighted MRI, C6–C7 axial image.

## Clinical Presentation

- A 55-year-old female presents with neck pain radiating into the interscapular region, and numbness in the left arm and hand.
- On examination, she is 5 feet 2 inches tall and weighs 275 pounds. Her motor function is intact. Sensation is diminished to light touch and pin prick in the left lateral three and half fingers. She has a positive Phalen's test on the left, as well as bilateral Tinel's sign over the median nerve at the wrist. She has a Hoffman's sign on the right side.
- Preoperative imaging is shown in ▶**Fig. 115.1**.
- Electromyography (EMG) reveals severe left and moderate right carpal tunnel syndrome.
- Selective left C7 epidural nerve root sleeve injection improved her interscapular pain but did not resolve subjective weakness in the left arm.

## Questions

1. What are the key findings on the MRI?
2. What does her clinical examination reveal?
3. How do you assess patients with cervical spondylotic myelopathy (CSM)? How do you counsel patients on the natural history of CSM?
4. In the case of a double crush syndrome, which pathology do you treat first?
5. What are your treatment options for CSM?
6. Would you offer her surgical treatment? If so, what procedure would you suggest?

## Answers

1. *What are the key findings on the MRI?*
   - MRI of the cervical spine reveals severe spinal stenosis from C4 to C7, predominantly caused by disc herniations, with increased T2 signal within the spinal cord.
   - In addition, there is an eccentric disc herniation at C6–C7 on the left, causing narrowing of the lateral recess and compression of the exiting C7 nerve root.
2. *What does her clinical examination reveal?*
   - Her examination is consistent with carpal tunnel syndrome, symptomatic on the left, with left C7 radiculopathy, without obvious signs or symptoms of myelopathy. (other than an abnormal Hoffman's sign)
3. *How do you assess patients with cervical spondylotic myelopathy (CSM)? How do you counsel patients on the natural history of CSM?*
   - Etiology of CSM is multifactorial. Degenerative disease processes cause loss of disc height, while posterior protrusion of the intervertebral discs and buckling of the ligamentum flavum ultimately lead to stenosis of the spinal canal and neural foramina.
   - Cervical spondylosis is subsequently the most common cause of cervical myelopathy.[1] Symptoms arise from compression of the spinal cord and/or its blood supply; patients may present with difficulty walking, imbalance, and clumsiness of the hands, among other signs and symptoms of upper motor neuron dysfunction.[2]
   - Studies by Fehlings et al found age, physical exam, duration of symptoms, EMG findings, and imaging to affect patients' outcomes.[3]
     - Patients younger than 75 years with mild to moderate CSM observed over a 3-year period were found to remain neurologically stable (class II evidence).[2,3]
     - For patients with cervical stenosis who are not yet myelopathic, EMG abnormality or clinically symptomatic radiculopathy was found to be predictive of developing myelopathy in the future (class I evidence).[3,4]
     - Patients with increased T2 signal within the spinal cord have worsened outcomes.
     - Duration of symptoms has a negative impact on overall outcomes, thus early treatment for symptomatic patients is advisable.
4. *In the case of a double crush syndrome, which pathology do you treat first?*
   - This is a controversial topic. "Double crush" is used to describe compressive pathology of the peripheral nerve in two anatomical locations along its course; in this case the left C7 nerve root and the left median nerve at the carpal tunnel.[5,6]
   - The decision of whether each pathology needs treatment, how to treat it, and in what sequence, should be carefully considered.
     - The argument to treat the carpal tunnel first: it is the simpler and faster procedure.
     - The argument to treat the myelopathy first: this is the more serious problem. In addition, patients with both cervical radiculopathy and carpal tunnel syndrome are more likely to view the carpal tunnel release as a failure due to persistent symptoms.[5]
     - Some surgeons may elect to address both issues in the same surgical setting, thus minimizing the recovery time for the patient and maximizing the outcome of surgery.
   - Due to our patient's radicular symptoms and the severity of stenosis, the cervical stenosis was treated initially, with bilateral carpal tunnel releases addressed at a later date.
5. *What are your treatment options for CSM?*
   - The main goal of CSM treatment is to decompress the cervical spinal cord and nerve roots. Other goals include restoring normal lordosis and stabilizing the cervical spine. Cheung et al found that 70% of patients undergoing decompression for CSM improved in regards to neurological function, with the greatest improvement in the arms, then legs, then sphincter function.[7] Patients tended to improve most significantly over the first 3 months postoperatively, and improvement plateaued by the 6-month mark.
     - Spinal cord decompression may be approached from anterior or posterior techniques based on the patient's pathology. Class III evidence shows no superiority of one approach compared with another, as long as the neural elements are decompressed[3] (see ▸**Table 115.1** for a summary).
     - Anterior approaches include multilevel anterior cervical discectomy and fusion (ACDF), with or without cervical corpectomy, used to address disc-based disease and/or anterior osteophytes.
       - Hillard et al found that anterior pathology without involvement posterior to the vertebral body should be addressed via multilevel discectomies. This provides greater control over kyphosis correction than cervical corpectomy, as distraction and fixation can be achieved at multiple levels. Cervical corpectomy, particularly if completed at multiple levels, leads to one large graft, which limits the gain in lordosis.[1]
     - Konig et al also approach cervical pathology with ACDF when this is enough to adequately address the anterior pathology. However, if the anterior pathology includes ossified posterior longitudinal

**Table 115.1** Advantages and disadvantages of anterior versus posterior approaches to the cervical spine

| Anterior Approach | Posterior Approach |
|---|---|
| **Advantages** | **Advantages** |
| • With adequate distraction, one can correct the kyp hotic deformity<br>• One can address anterior pathology directly such as discs and bony spurs<br>• Sufficient distraction can unbuckle the ligamentum flavum and indirectly treat a posterior pathology<br>• It is typically better tolerated in patients with 1 or 2 level disease and younger patients | • Posterior approaches are usually technically easier for neurosurgeons, therefore more suitable for older and osteoporotic patients<br>• There are virtually no risks to the cranial nerves and carotid arteries (i.e., preferred procedure for professional singers)<br>• One can address posterior pathology such as ligamentum flavum buckling or ossification directly<br>• There is a lower risk of incidental durotomy in cases of OPLL |
| **Disadvantages** | **Disadvantages** |
| • There is a risk of cranial nerve palsy with lower level disease (dysphagia, dysphonia) and a greater risk of vascular and esophageal injury<br>• It requires bony fusion and the associated morbidity of bony fusion<br>• There is a high risk of subsidence in osteoporotic patients<br>• There is a lesser likelihood of fusion in greater than 2-level construct cases, unless it is supplemented with a posterior approach procedure | • There is a greater risk of kyphotic deformities or instability in pure laminectomy cases (if too much facet is taken)<br>• There is an inability to directly address anterior pathology<br>• More pain is usually experienced postoperatively along the paraspinal muscles especially in younger patients<br>• C5 nerve root palsy is described more often with posterior decompressions |

## Answers (*continued*)

ligament (OPLL), caudal or rostral extension of disc material, or significant posterior osteophyte complexes, cervical corpectomy may be used in isolation or in addition to multilevel discectomies.[8] Posterior approaches including laminoplasty or laminectomy, with or without posterior cervical fusion, are employed when the primary pathology is posterior in nature, such as ligamental and/or facet hypertrophy.

  - ○ Laminoplasty aims to decompress the spine without causing future kyphosis, which is a known complication in 10 to 45% of cervical laminectomy cases.[2] Laminoplasty is frequently used to treat diffuse OPLL, cervical stenosis in a straight or lordotic cervical spine, or to provide access to spinal tumors. Multiple variations on surgical technique exist. Hinge procedures, also called “open-door” laminoplasty, completely disconnect the lamina from the lateral mass on one side. A partial thickness trough is drilled on the opposite side and used as a fulcrum to swing the posterior elements dorsally. Wires or laminar plates and screws are used to keep the posterior elements in place.[9] The maximum decompression of neural elements is seen at the side of opening in “hinge” procedures. Alternatively, “French door” procedures allow for symmetric expansion of the canal. Partial troughs are drilled bilaterally at the laminar/facet junction, while the spinous process is split completely. Each side is then hinged open and secured with suture, wiring or plates, and screws.[9] Most importantly, patients’ neurological recovery rates ranged from 50 to 70%, independent of the surgical technique used.[9]
  - ○ Clinically, outcomes of laminectomy patients are similar to those undergoing ACDF in the short term, but there is a risk for kyphosis in the long term.[3] It has also been argued that posterior fusion leads to improved neck pain as compared with nonfusion techniques.[8]
  - ○ Patients with fixed kyphotic deformities are not appropriate for posterior decompression procedures, and should have anterior surgical approaches considered.[1,2,8]

– Alternatively, circumferential decompression and fusion may be indicated for patients with severe kyphotic deformity, evidence of instability or systemic concerns for poor fusion quality.[8] Multicolumn injury from trauma may also require anterior and posterior surgical techniques in order to achieve adequate stabilization, reduce postoperative bracing requirements (in order to allow faster rehabilitation), or to completely achieve decompression and realignment.[10] Furthermore, patients with post laminectomy kyphosis are likely to fail anterior fusions due to the lack of posterior tension band; combined circumferential approaches are indicated for these patients. Surgeons may choose to complete circumferential cases in a single setting or as a staged procedure based on the patient’s tolerance for prolonged anesthesia.[10] The more complicated cases with multiple prior surgeries and a fixed kyphotic deformity may

## Answers (*continued*)

even require a back-front-back or a front-back-front operation.

6. *Would you offer her surgical treatment? If so, what procedure would you suggest?*
   - Cervical stenosis without myelopathy can be carefully observed. Patients remain at increased risk for potential spinal cord injury with even minor trauma. In the presence of severe stenosis, signal changes in the spinal cord, and some signs and symptoms of myelopathy, this patient should be offered elective surgery.
     - Her radicular symptoms may be addressed surgically given only short-term relief with steroid injection. If lateral recess narrowing were her only pathology, this could be treated with a posterior foraminotomy. Given her extensive cervical stenosis, it would be prudent to consider decompression of the spinal cord as well as the symptomatic nerve root.
     - Her pathology is anterior, due to disc degeneration and herniation. Based on her imaging, this could be adequately treated from multilevel discectomies and fusion. Not only will this approach sufficiently decompress the spinal cord, it will also restore cervical lordosis. Placement of grafts at each level also helps to distract the cervical spine, which can be used for correction of ligamentum flavum buckling (not seen in this case). While she is obese, she did not have significant past medical history that would increase her risk of pseudoarthrosis, thus a combined anterior-posterior approach was not felt to be necessary. She underwent a C4–C7 ACDF and did well in recovery; see postoperative imaging in ▸**Fig. 115.2**. Eighteen months post ACDF, she underwent carpal tunnel release for the left hand, followed by the right hand 3 months later. She continues to do well postoperatively at 2.5 years, with resolution of her left arm symptoms.

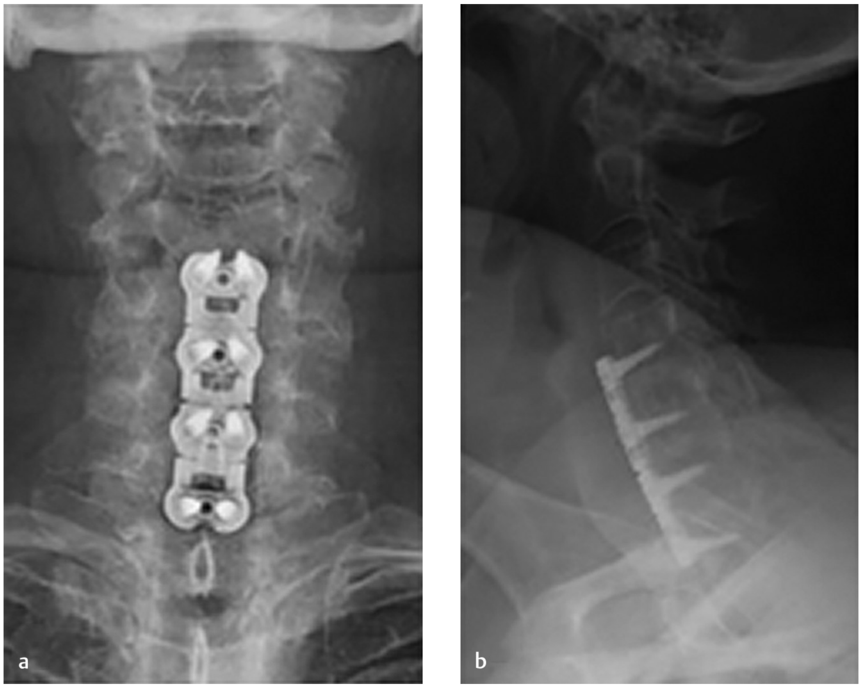

**Fig. 115.2** Postoperative imaging. (**a**) Cervical spinal X-ray, anteroposterior; (**b**) Cervical spinal X-ray, lateral.

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# Case 116 Ossification of the Posterior Longitudinal Ligament

*Justine Pearl, Anup Aggarwal, and Remi Nader*

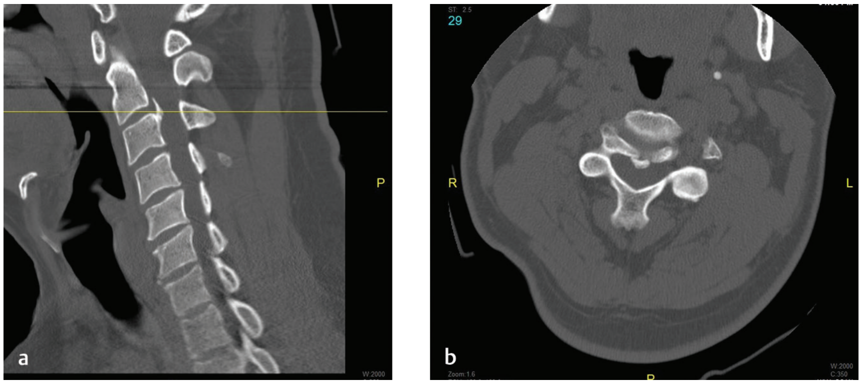

**Fig. 116.1** (**a**) CT sagittal view of the cervical spine. (**b**) CT axial view of the cervical spine through C3–C4.

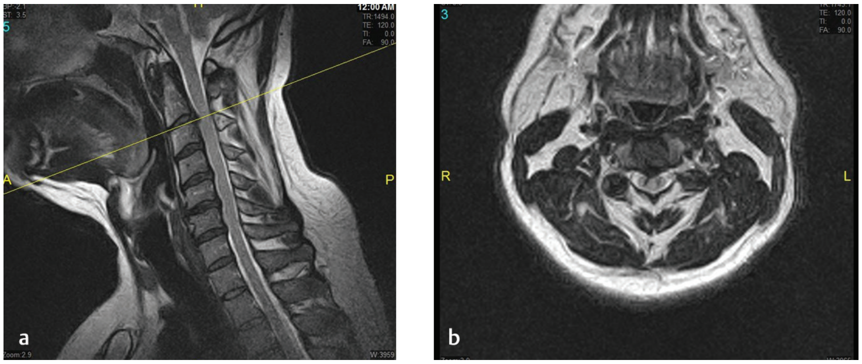

**Fig. 116.2** (**a**) MRI T2-weighted image, sagittal view of the cervical spine. (**b**) MRI T2-weighted image, axial view of the cervical spine through C3–C4.

## Clinical Presentation

- A 45-year-old man presents to the emergency room with neck pain that radiates to right shoulder. He describes pain as stabbing and is rated 6/10 on a visual analogue scale. It started when he was involved in a motor vehicle accident one day prior. Associated symptoms include numbness at the base of neck and stiffness, headaches, and light sensitivity.
- The neurosurgery service is consulted due to findings on the CT (▶**Fig. 116.1**) of the cervical spine. His physical examination reveals normal strength and sensation to light touch and pinprick. He is hyperreflexic in the upper and lower limbs. Hoffman's reflex is negative bilaterally and he is not frankly myelopathic. There is no perianal anesthesia and digital rectal examination is normal for tone.
- MRI is obtained (▶**Fig. 116.2**).

## Questions

1. Interpret the CT and MRI images (▶**Fig. 116.1**, ▶**Fig. 116.2**).
2. Describe the anatomy of the posterior longitudinal ligament (PLL).
3. Is the morphology of the ligament constant throughout the length of the spine?
4. How does the PLL become “ossified”?
5. What is the etiology of ossified PLL (OPLL)?
6. What is the incidence of OPLL? Is there a difference between males and females?
7. What other associated conditions may present with OPLL?
8. What is the rate of growth of ossification of the PLL?
9. Classify OPLL.

## Questions (*continued*)

10. What is the most common distribution of OPLL throughout the spine?
11. Describe the pathophysiology of OPLL's effects on the spinal cord.
12. What conservative options are available to offer the patient?
13. What factors affect outcome in patients managed surgically?
14. What are the indications for surgery, and what are the surgical options?

The patient undergoes an anterior approach for resection of OPLL with discectomy and fusion. Intraoperatively at the C3–C4 level as the calcified portion of the ligament is attempted to be resected, a large gap in the dura is visualized and the patient has a wide opening in the thecal sac with cerebrospinal fluid (CSF) leakage.

15. How do you manage this complication?

## Answers

1. *Interpret the CT and MRI images (*▶**Fig. 116.1**, ▶**Fig. 116.2***).*
   - CT scan shows that there is cervical spondylosis from C3 through C6. The PLL is ossified at the C3–C4 level.
   - MRI T2 sagittal sequence reveals focal OPLL at the C3–C4 level. There is multilevel hypertrophic facet disease. There are multiple levels of disc bulges, specifically at C3–C4, C4–C5, C5–C6, and C6–C7. There is also multilevel moderate spinal canal stenosis. There is some loss of lordosis, however alignment is maintained.
2. *Describe the anatomy of the posterior longitudinal ligament (PLL).*
   - The PLL originates from the basiocciput and lies along the posterior aspect of the vertebral bodies. Its insertion point is the posterior margin of the sacrum.
   - There are segmental attachments to the annulus of each disc and the adjacent vertebral body cortex.
   - At the midvertebral body, the area where the basivertebral venous plexus lies separates the PLL from the rest of the bony cortex.
3. *Is the morphology of the ligament constant throughout the length of the spine?*
   - No. In the cervical spine region, the ligament should be uniformly broad and flat.
   - At the lower levels of the spine, the PLL is quite contracted in the midline and then expands laterally.
4. *How does the PLL become "ossified"?*
   - Mechanical stress on the ligament initiates the process. This results in hypervascular fibrosis and ligamentous hypertrophy, followed by mineralization of the ligament. This is especially notable in the portion of the PLL fibrously attached to the vertebral body.
   - The cartilage cells in the vertebral body's periosteum proliferate, and this is followed by cartilage cell proliferation within the annulus. The dura and the PLL are then affected, with cartilage cell proliferation and calcification by endochondral ossification (*the key process in the formation of OPLL*). Finally, mature lamellar bone is formed with Haversian canals.
5. *What is the etiology of ossified PLL (OPLL)?*
   - Many causative factors have been implicated in the disease. Fluoride intoxication, diabetes mellitus, growth hormone imbalance, disc herniations, recurrent minor trauma, abnormal calcium metabolism, and infection have all been suggested.
6. *What is the incidence of OPLL? Is there a difference between males and females?*
   - Incidence in the Japanese/Asian population: 2.4%
   - Incidence in Caucasians: 0.2 to 0.7%
   - Males are more affected than females.
7. *What other associated conditions may present with OPLL?*
   - Diffuse idiopathic skeletal hyperostosis (DISH)
   - Ossification of the ligamentum flavum
   - Ankylosing spondylitis
   - Schizophrenia: possibly
8. *What is the rate of growth of ossification of the PLL?*
   - This varies and there are no clear numbers: this is dependent on patient and genetic factors.
   - Generally, the OPLL will compress the canal by roughly 0.4 mm/year, and longitudinal growth is about 0.7 mm/year.
9. *Classify OPLL.*
   - The most commonly used classification is that of Hirabayashi and is as follows (see ▶**Fig. 116.3**):
     - Segmental: OPLL behind each vertebral body (39%)
     - Continuous: (27%)
     - Localized: OPLL over the intervertebral disc space (8%)
     - Mixed: (29%)

     OPLL is most frequently seen at C4, C5, and C6, and the average number of vertebral bodies involved is three. It is relevant to note that the ratio of maximal thickness of OPLL to the anteroposterior diameter of the spinal canal is highest in the mixed and continuous types.

## Answers (*continued*)

10. *What is the most common distribution of OPLL throughout the spine?*
    - Cervical: 70% (especially C3–C4)
    - Thoracic: 15% (especially T4–T6)
    - Lumbar: 10% (especially L1–L3)
11. *Describe the pathophysiology of OPLL's effects on the spinal cord.*
    - Stages of spinal cord damage by OPLL are as follows, graded 0 to 3:
      - 0: normal or mild compression of anterior horns without neuronal loss
      - 1: mild compression of anterior horns with partial neuronal loss
      - 2: marked deformity of anterior horn with severe neuronal loss
      - 3: severe spinal cord damage with cystic loss/myelomalacia
    - The most significant destruction is in the central gray matter of the cord. Tissue necrosis and central gray matter cavitation is evident with seriously damaged cord and can extend to the more ventral portion of the posterior columns.
    - Anatomically, this corresponds to the so-called watershed zone of the cord in the boundary area between the anterior and posterior spinal artery supply, where ischemia and/or venous stasis may be present.
    - In autopsy studies, anterior horn cells can be reduced in both number and size. Demyelination changes can also be seen in the white matter.
12. *What conservative options are available to offer the patient?*
    - Conservative management aims to relieve irritation but do not address a patient's myelopathy directly. Nonsteroidal anti-inflammatory drugs, traction, limited bed rest, and halter traction are some options. Continuous skull traction with halo to eliminate dynamic motion (which further irritates the ligament and fosters the ossification cycle) has also been proposed.
    - Generally, in patients with only a mild disturbance in their activities of daily living, follow-up after 5 years of conservative treatment showed that 55% of patients remain stable, 27% improved, and 19% had worsening of their symptoms.
13. *What factors affect outcome in patients managed surgically?*
    - Multiple factors impact outcomes in the OPLL patient. These include the degree of disability of the patient in their activities of daily living, presence of comorbid medical conditions, the duration and degree of myelopathy, the patient's age, any history of trauma, degree of preoperative spinal canal stenosis, and the degree of kyphosis of the spine.
14. *What are the indications for surgery, and what are the surgical options?*
    - Japanese Orthopedic Association (JOA) score of 6 to 12 points
    - Young patients with severe spinal stenosis, even if their myelopathy is not severe, they should be considered for surgery
    - Pain, especially radiating to the upper arm, refractory to medical management
    - Progressive symptoms and/or progressive myelopathy
    - Some authors advocate operative management if there is substantial spinal cord compression or signal change within the cord on T2-weighted MRI.

      **Options:**

      OPLL can be managed via both an anterior, posterior, or combined approach.
    - Anterior options:
      - Anterior vertebrectomy and interbody fusion with instrumentation
        - ○ Generally considered when patient is < 65 years old, or older than 65 years but with kyphosis.
        - ○ In some areas, the dura and PLL are indistinguishable
        - ○ Postoperative halo can be used if more than two levels of vertebrectomy are performed, or one can also consider supplementing this with a posterior fusion/fixation.
        - ○ The ossified ligaments compressing the spinal cord are either extirpated or are floated.
        - ○ In the segmental and localized types of OPLL, below the C3–C4 level, anterior decompression and vertebral body fusion is generally the preferred approach when fewer than three disc levels are affected. This approach produces particularly good results for patients with clumsiness of the fingers, severe intrinsic muscular atrophy caused by lesions of the anterior horn cells, or both. It is, however, a more extensive and risky surgery than a posterior decompression. Postoperatively, 85 to 90% will show neurological improvement.
      - Major risks include:
        - ○ Worsened myelopathy (2–20% incidence)
        - ○ Radiculopathy, especially at the C5 level (15%)
        - ○ CSF leak (15–25%)
        - ○ Graft extrusion, nonfusion
        - ○ Anterior approach standard risks: esophageal injury, Horner's syndrome, hoarseness, vertebral artery injury
      - Anterior floating method with an autograft fusion
        - ○ In this procedure, a thin layer of OPLL adherent to the dura is intentionally left behind during corpectomy.

## Answers (*continued*)

    - ○ The goal is to avoid CSF leak and cord injury.
    - ○ Indications and risk profile are otherwise similar to above method.
- Posterior options:
  Posterior approach surgery does not address the compressive pathology directly, as the issue is the anteriorly compressed cord by the OPLL mass, but it is still an option in the medically unfit patient or the severely degenerative spine.
  - – Laminectomy
    - ○ If three or more levels are affected, this is a reasonable approach for an older patient without significant kyphosis.
    - ○ There are significant risks associated with laminectomy alone for the treatment of OPLL, including but not limited to spinal cord injury during decompression, radiculopathy (especially at C5), recurrence/progression of myelopathy secondary to the persistence of or further growth of the lesion, and development of a swan-neck kyphotic deformity.
    - ○ In the continuous and mixed types of OPLL, the ossified ligaments may be biologically stimulated by the operative procedure, accelerating the postoperative growth of the OPLL mass. In addition, any structural weakness caused by a laminectomy alone may cause postoperative growth of the OPLL mass in a compensatory process not unlike how the initial mass was formed.
  - – Laminoplasty
    - ○ Not a viable option if any kyphotic deformity is present
    - ○ Sixty to 70% will improve neurologically
    - ○ Can help maintain lordosis and spinal column stability
    - ○ Similar risks as laminectomy, but higher incidence of postoperative C5 radiculopathy (5–10%)
  - – Advantages:
    - ○ Technically easier and safer
    - ○ Postoperative support/stability is better than laminectomy
    - ○ Dynamic forces that may result in further PLL damage can be significantly reduced or eliminated, since the range of motion of the preoperative cervical spine will be reduced by 50%, by ankylosing of the bony gutter on the hinged side of the construct.
  - – Complications:
    - ○ Closure of the opened laminae. Adding "stay" sutures between spinous processes and facet capsules may help keep the construct "open."
    - ○ Transient muscle paraparesis of the shoulder girdle and severe neck pain: results from a tethering effect of the nerve roots of C5 or C6.
  - – Laminectomy and lateral mass fusion and instrumentation
    - ○ In the continuous type of OPLL, laminectomy and expansive laminoplasty for posterior decompression can be performed.
    - ○ Posterior decompression should extend as far as one level below and above the stenotic site.
    - ○ Nerve root decompression can also be done in the same surgery
    - ○ Prevents the future risk of kyphosis and OPLL expansion as seen with laminectomy alone, however more technically demanding than a simple laminectomy
    - ○ Added risk of posterior instrumentation: for example, hardware failure, pseudoarthrosis, vertebral artery injury, or nerve root injury during lateral mass screw placement.

15. *How do you manage this complication?*
    - There are multiple options to manage a dural defect with CSF leakage from an anterior approach. The goals should include sealing off the defect and avoiding any CSF seepage through the skin. One should also aim for minimizing the chances of development of a pseudomeningocele postoperatively which may potentially compromise upper airway and laryngeal structures.
    - Harvesting some fascia, muscle, and/or fat to seal off the defect as a primary option can be attempted. This, with the addition of fibrin glue material such as Tisseel (Baxter International, Deerfield, Ill) can act as a sealant along the anterior dura. It is often difficult to suture a defect in the anterior dura. Another option is to use synthetic material in order to close that defect such as type 1 collagen matrix (DuraGen, Integra LifeSciences, Plainsboro, NJ) (or other similar collagen material).
    - One should also strongly consider placing a lumbar drain for 3 to 5 days and monitoring the patient in a close monitoring unit so as to avoid overdrainage (up to 10–15 cc of CSF per hour). The patient's head should be kept elevated postoperatively at ~30 degrees if not higher and the patient should be monitored for the formation of pseudomeningocele. Antibiotics should be used if a lumbar drain is placed. In most cases by placing an anterior grafting and a lumbar drain, the problem should be well contained.
    - In occasional cases when these measures are not sufficient and there is still seepage of CSF or formation of symptomatic pseudomeningocele, one should consider a second look surgery with preoperative CT/MRI and planning with a plastic surgeon or a head and neck surgeon for the possibility of a myofascial flap rotation or further grafting as necessary.

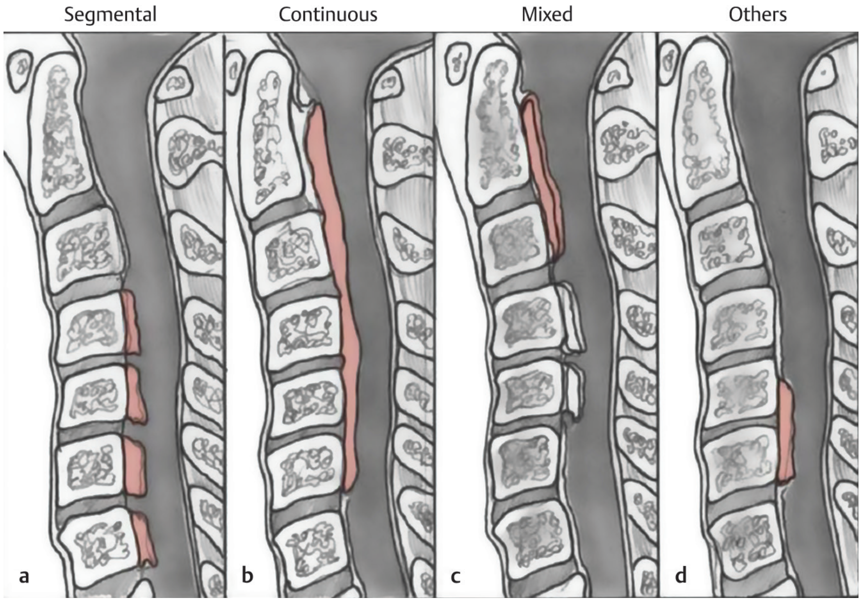

**Fig. 116.3** Hirabayashi's OPLL classification: (**a**) Segmental; (**b**) Continuous: from body to body; (**c**) Mixed: features of both segmental and continuous; (**d**) Other: confined to the disc space. (Reproduced with permission from Nader R., et al. Neurosurgery Tricks of the Trade—Spine and Peripheral Nerve. New York: Thieme; 2013.)

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# Case 117 Thoracic Disc Herniation

*Remi Nader and Mohammad Almubaslat*

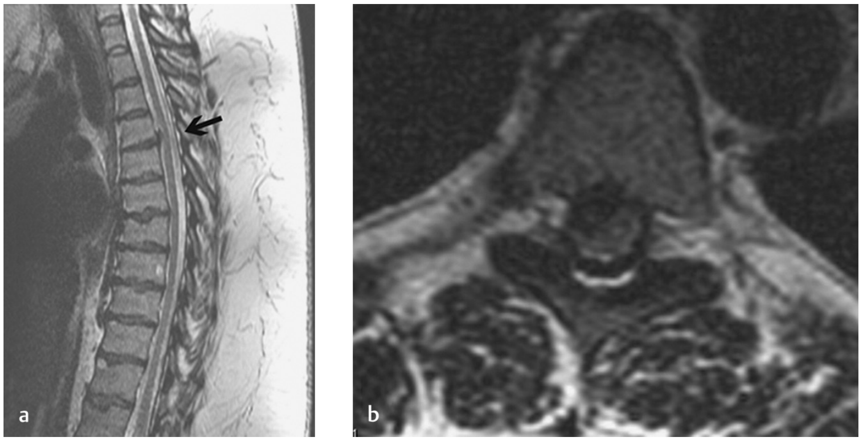

**Fig. 117.1** (**a**) Sagittal and (**b**) axial T2-weighted MR images of the thoracic spine showing herniated disc (*arrow*) at the T4–T5 level.

## Clinical Presentation

- A 31-year-old woman presents with a 3-month history of mid-to-lower-back pain radiating to both hips. The pain starts in the midscapular area.
- She also suffers from progressive balance problems; she uses a walker and falls frequently.
- She describes her legs as feeling disconnected from the rest of her body.
- Examination reveals a morbidly obese woman, mild weakness in both iliopsoas muscles, sensory level at T4, lower extremity hyperreflexia, bilateral clonus, and a severely spastic gait.

## Questions

1. What is your differential diagnosis?
2. What studies would you like to obtain?

   MRI of the cervical spine is read as normal by the radiologist. You then decide to obtain an MRI of the thoracic and lumbar spine. The thoracic spine MRI is shown in ▶**Fig. 117.1**. The lumbar MRI is read as normal.

3. Give a general classification of thoracic disc herniations.
4. What are the different treatment measures?
5. Which one would you select for this patient and why?
6. What are the different surgical approaches to a thoracic disc, their advantages and limitations?
7. Which approach would you select in this case?
8. What are the general outcomes of each approach?
9. What are the indications for fusion after discectomy?

## Answers

1. *What is your differential diagnosis?*
   - The differential diagnosis of spinal progressive myelopathy is broad and includes the following[1,2] (for a more detailed diagnosis list, please refer to Case 87, Postherpetic Neuralgia):
     - Syringomyelia
     - Postviral conditions
     - Vertebral osteomyelitis
     - Traumatic fracture
     - Spinal cord tumor
     - Epidural lipomatosis
     - Degenerative conditions: spondylotic myelopathy, ossified posterior longitudinal ligament, disc herniation
     - Multiple sclerosis, transverse myelitis, or Guillain–Barré syndrome
     - Spinal epidural, subdural, or subarachnoid hematoma, spinal cord infarction
2. *What studies would you like to obtain?*
   - Imaging studies
     - Plain radiographs
     - MRI
       - Study of choice
       - Noninvasive
       - >95% effective in diagnosis of myelopathy
     - CT and/or CT myelogram
       - When MRI cannot be done
       - In cases where resolution of MRI is low
       - Plain CT good for C5–C6 but poor for C6–C7 or below due to shoulder artifact
       - CT myelogram has 98% accuracy but is more invasive and may require overnight admission
       - In thoracic disc herniations, CT scan helps to evaluate the extent of calcifications.
3. *Give a general classification of thoracic disc herniations.*
   - Classification by level[3,4]
     - T1–T4, or upper thoracic spine
       - Thoracic outlet and superior mediastinum
     - T5–T9, or middle thoracic spine
       - Stabilizing influences of the rib cage
     - T10–T12, or thoracolumbar spine
       - Transition zone of vertebral configuration
   - Classification by laterality[4]
       - Midline
       - Paramedian
       - Lateral
4. *What are the different treatment measures?*
   - Conservative measures[4]
     - Analgesics, muscle relaxants, nonsteroidal anti-inflammatory drugs
     - Limited activity, bed rest for a few days, postural modifications and re-education
     - Physical therapy: extension exercises and paraspinal muscle strengthening
     - Stretching and muscle relaxation with heat, ultrasound, or transcutaneous electrical stimulation
     - Bracing (thoracolumbar spinal orthosis brace)
     - Short course of oral steroids
     - Epidural steroid injections facet blocks, median branch blocks
   - Surgical measures—indications[4,5]
     - Failure of conservative measures
     - Severe symptoms of pain or myelopathy
     - Progressive deterioration in symptoms
5. *Which one would you select for this patient and why?*
   - Surgical treatment is the most appropriate approach in this case.
   - This is due to the severity of the symptoms and signs of myelopathy.
6. *What are the different surgical approaches to a thoracic disc, their advantages and limitations?*
   Approaches for thoracic disc herniations[4–9]
   - Posterior midline laminectomy
     - Advantages:
       - Ease and familiarity of anatomy and approach
     - Disadvantages:
       - Unacceptably high failure rate in single-level anterior pathology
       - Neural injury
       - Inadequate decompression
       - *Not* a generally accepted approach for most thoracic discs
   - Posterolateral approach: transpedicular or transfacet—pedicle sparing
     - Advantages:
       - Laminectomy, removal of pedicle and medial facetectomy
       - Good results for lateral herniated disc
       - Do not require transpleural or transmediastinal dissection
       - No violation of the rib cage or ligation of the neurovascular bundle
       - Potential for less operative time and bleeding
       - Familiar to most neurosurgeons
       - May be done endoscopically
     - Disadvantages:
       - Limited visibility of the midline of the anterior spinal canal
       - Unable to completely remove discs that are mainly central or past the midline
       - Possible instability with removal of pedicle/facet complex
   - Costotransversectomy
     - Advantages:
       - Good for lateral disc herniation
       - Resection of transverse process and 4 to 8 cm of rib: rib that articulates with the inferior vertebra of the disc level to be treated

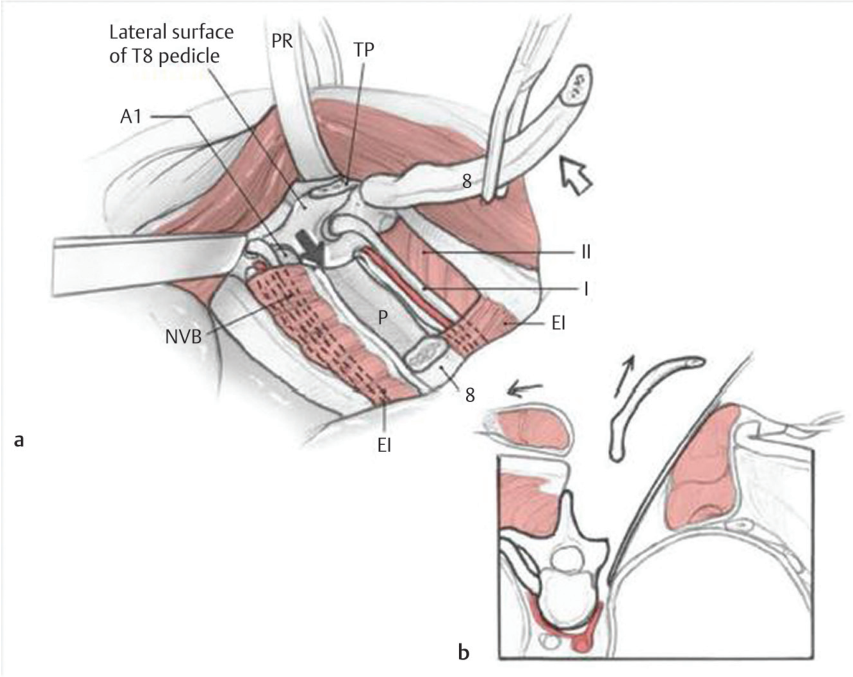

**Fig. 117.2** Artist's rendering of the lateral extracavitary approach: parietal pleura and contents retracted to expose amputated left T8 transverse process and pedicle, the T7–T8 annulus, and the foramen with exiting nerve roots at these levels. The retractor point to the segmental vessels along the lateral wall of T8 vertebral body. (A1, T7/8 disc annulus; NVB, T7 neurovascular bundle, PR, Penrose drain, TP, transverse process; thick arrow, segmental vessels along T8 body; EI, external intercostals muscle). (Reproduced with permission from Nader R., et al. Neurosurgery Tricks of the Trade—Spine and Peripheral Nerve. New York: Thieme; 2013.)

## Answers (*continued*)

        - Ligation of intercostal nerve ~3 cm distal to dorsal root ganglion
        - Direct visualization of neural elements
    - Disadvantages:
        - Risk of interrupting radicular artery which supplies blood to spinal cord
        - Risk of intercostal neuralgia or anesthesia dolorosa
        - Manipulation of intercostals muscles
- Lateral extracavitary approach (see ▶**Fig. 117.2**)
    - Advantages:
        - Enables total resection of centrally located discs
        - Good for central soft and calcified discs
        - Ease of multilevel exposure
        - No need for chest tube drainage if pleura preserved
    - Disadvantages:
        - Extensive procedures and bony resection
        - High operating times, blood loss
        - Significant perioperative pain and physiologic stress to the patient
- Anterolateral transthoracic approach
    - Advantages:
        - Good for central disc or when myelopathy is present → best operative results
        - Good exposure obtained from T4–T5 to T11–T12
        - Right-sided thoracotomy preferred for midthoracic because heart does impede access
        - Left-sided preferred for lower thoracic (easier to mobilize aorta than vena cava)
        - Low risk of mechanical cord injury
        - Little compromise of stability
    - Disadvantages:
        - Requires thoracic surgeon
        - Extensive, time-demanding procedure
        - Risk of vascular cord injury
        - Pulmonary complications and cerebrospinal fluid (CSF) pleural fistula
        - Postthoracotomy pain syndrome
        - Closed chest drainage is required postoperatively.
        - Mediastinal structures are at increased risk.
        - Hard to access discs above T4–T5
- Video-assisted thoracoscopic approach
    - Advantages:
        - Minimally invasive
        - Avoids pulmonary complications and morbidities
        - Less blood loss
        - Can access centrally located disc herniations
        - Prevents denervation of paraspinal musculature
        - Less bone resection
    - Disadvantages:
        - Requires thoracic surgeon
        - High level of technical skill and steep learning curve
        - Limited ability to strut graft or fuse anteriorly
        - Difficult to repair CSF leak

7. *Which approach would you select in this case?*
    - Any of the approaches described above are appropriate except for laminectomy.
    - In this case, a posterolateral approach via costotransversectomy was selected. Postoperative MRI is shown in ▶**Fig. 117.3** and ▶**Fig. 117.4** for illustration of the approach.

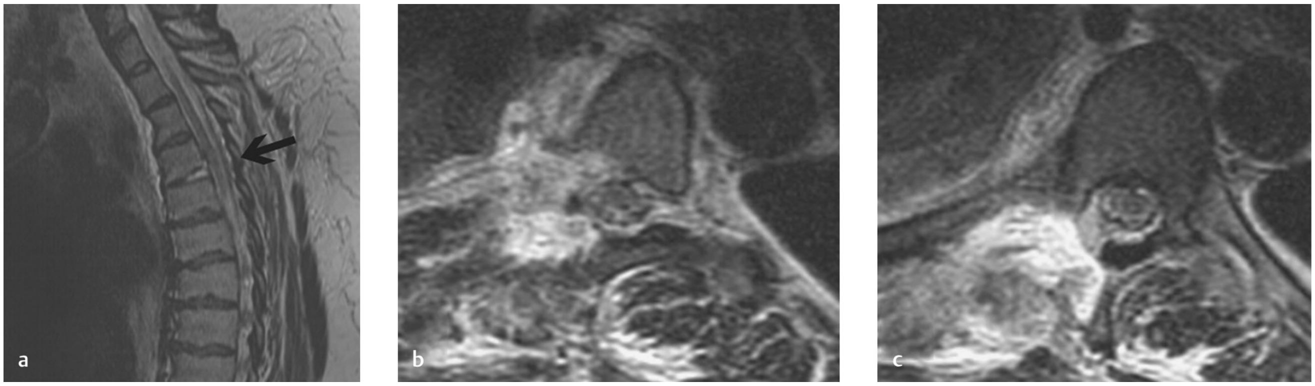

**Fig. 117.3** (**a**) Sagittal and (**b, c**) axial T2-weighted MR images of the thoracic spine showing herniated disc resection (*arrow*) at the T4–T5 level via a costotransversectomy approach.

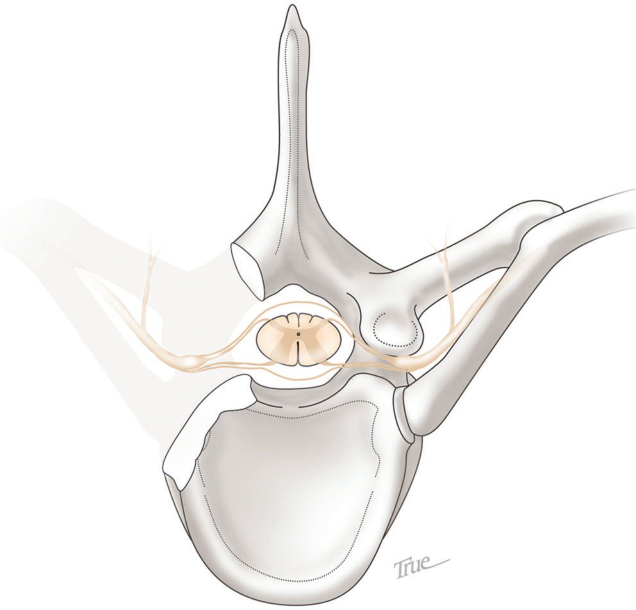

**Fig. 117.4** Artist's rendering of the costotransversectomy approach, showing the maximal extent of bony removal. This includes part of the vertebral body, the lamina, transverse process, and pars interarticularis. (Reproduced with permission from Wolfla CE, Resnick DK. Neurosurgical Operative Atlas. Spine and Peripheral Nerves. New York: Thieme Medical Publishers/American Association of Neurological Surgeons; 2007:151.)

## Answers (*continued*)

- The reasoning behind this selection: the patient is young and the disc is more likely noncalcified and the disc is lateral to paramedian.

8. *What are the general outcomes of each approach?*
   - Overall outcomes[9–12]
     - Posterior midline laminectomy: 32% worse outcome, 57% improved or stayed the same
     - Posterolateral approach: 7% worse outcome, 82% improved or stayed the same
     - Endoscopic transpedicular: 90% improvement ($n = 25$)[9]
     - Costotransversectomy: 0% worse, 12% same, 88% improved
     - Anterolateral transthoracic approach: 0% worse, 94% pain improved, 97% myelopathy improvement
9. *What are the indications for fusion after discectomy?*
   - Indications for fusion[4]
     - Multilevel disc resection
     - Kyphosis
     - Wide vertebral body segment resection affecting stability
     - Junctional level (T12–L1)

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# Case 118 Thoracolumbar Scoliosis

*Sunil Kukreja and H. Francis Farhadi*

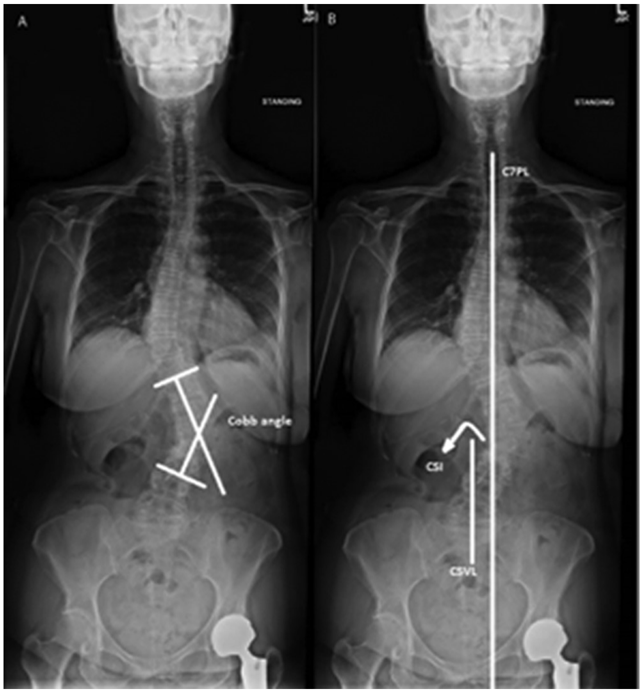

**Fig. 118.1** Full-spine standing anteroposterior X-rays showing measurement of parameters in the coronal plane.

## Clinical Presentation

- A 62-year-old female presents with worsening midthoracic and lower back pain over 5 years.
- The back pain is exacerbated by either sitting or standing for extended periods.
- She also complains of associated radicular pain of the left lower extremity.
- On exam: muscle strength is 5/5 all groups; there were no sensory deficits; a scoliotic deformity of the thoracolumbar spine was noted.
- Anteroposterior (AP) and lateral standing X-rays of the spine were obtained (▶**Fig. 118.1** and ▶**Fig. 118.2**).

## Questions

1. Describe the X-ray findings.
2. What radiological parameters will you measure? Describe the methodology.
3. Which additional radiological images will you obtain and why?
4. How will you classify this deformity?
5. What are the indications for and the goals of surgery?
6. Describe the surgical options for deformity correction.
7. Describe the complications associated with adult spinal deformity (ASD) surgery.
8. What are the health-related quality of life (HRQOL) measures and instruments utilized to assess functional outcomes?
9. What are the radiological factors that best correlate with functional outcomes?

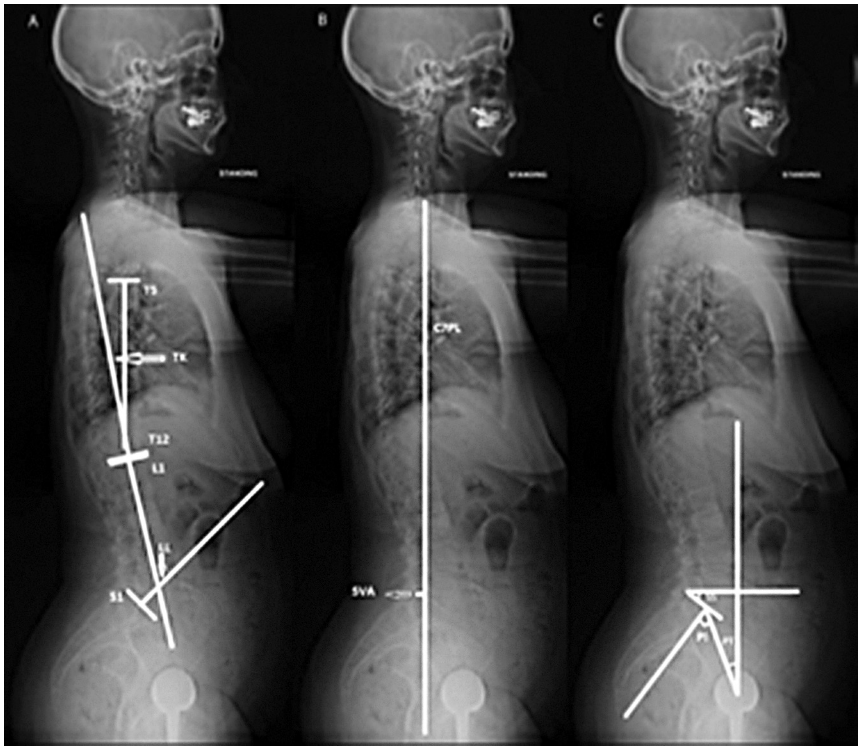

**Fig. 118.2** Full-spine standing lateral X-rays showing measurement of parameters in the sagittal plane.

## Answers

1. *Describe the X-ray findings.*
   - The AP full spine standing view (▶**Fig. 118.1**) reveals levoscoliosis of the lumbar spine from T12 to L4 with the apex at the L2 vertebral body. There is associated dextroscoliosis of the thoracic spine extending from T5 to T12.
   - The scoliotic curves are well compensated with no discrepancy at the shoulder level.
   - The lateral view (▶**Fig. 118.2**) shows relative loss of lumbar lordosis (LL) and kyphosis at the thoracolumbar (TL) junction from T11 to L2. There is compensatory reduction of thoracic kyphosis (TK) and increase in pelvic retroversion.
2. *What radiological parameters will you measure? Describe the methodology.*
   - The following radiological parameters will be measured:
     - Coronal: Cobb (CC) angles, convexity of the curve, degree of apical vertebra rotation (by Nash–Moe method), extent of the curves, coronal spinal imbalance (CSI).
     - Sagittal: TK, LL, TL kyphosis, sagittal vertical axis (SVA) and pelvic parameters including sacral slope (SS), pelvis tilt (PT), and pelvic incidence (PI).
   - Methods:
     - CC angle: We choose the most tilted vertebrae above and below the apex of the curve. The angle between intersecting lines drawn perpendicular to the lines drawn along the superior endplate of rostral vertebra and inferior endplate of caudal vertebra is the Cobb angle (▶**Fig. 118.1**a).
     - CSI: The distance between C7 plumb line (C7PL) and central sacral vertical line (CSVL) in the coronal plane (▶**Fig. 118.1**b).
     - The Nash–Moe method is a radiological measurement of vertebral rotation. The degree of rotation is calculated by measuring the percentage displacement of the convex pedicle with respect to the vertebral body width (▶**Fig. 118.3**).
     - TK: The Cobb angle measured from the superior end plate of T5 to the inferior endplate of T12 on the sagittal full-length film (▶**Fig. 118.2**a).
     - LL: The Cobb angle measured from the superior end plate of L1 to the inferior endplate of S1 on the sagittal full-length film (▶**Fig. 118.2**a).
     - TL kyphosis: The Cobb angle measured from the superior end plate of T10 to the inferior endplate of L2 on the sagittal full-length film.
     - SVA: A vertical plumb line drawn from the center of the C7 vertebral body. The plumb line is normally within 5 cm of the posterosuperior corner of the S1 endplate (▶**Fig. 118.2**b).
     - PI: The PI is a nonpositional anatomical parameter defined as the angle between the line perpendicular to the sacral plate at its midpoint and the line connecting this point to the axis of rotation of the femoral head (▶**Fig. 118.2**c).
     - SS: The SS is a positional parameter defined as the angle between the superior endplate of S1 and a horizontal line extending from the anterior-inferior corner of the S1 endplate (▶**Fig. 118.2**c).

## Answers (*continued*)

- PT: The PT is a positional parameter defined as the angle between the line connecting the midpoint of the superior sacral end plate to the femoral rotational axis and the line extending vertically from the femoral rotational axis (▶**Fig. 118.2**c). The radiological parameters for this patient include the following: CC = 44 degree, CSI = 1.8 cm, TK = 14 degree, LL = 48 degree, TLK = 11 degree, PI = 77 degree, SS = 52 degree, PT = 25 degree, PI–LL mismatch = 29 degree. (see ▶**Fig. 118.5**).

3. *Which additional radiological images will you obtain and why?*
   - We will also obtain dynamic views (lateral flexion-extension and right-left bending views). The flexion-extension films will identify any associated segmental instability in the sagittal plane. The flexibility of the coronal curves is evaluated with the bending films.
   - Standard CT and MRI scan are performed to further evaluate the bony and neural elements.
4. *How will you classify this deformity?*
   - Several classification systems are used to describe ASD. The Scoliosis Research Society (SRS) Schwab scale is a recent addition (published in 2012) that incorporates both coronal and sagittal radiological parameters. It consists of four components: coronal curve type and three sagittal modifiers (PI–LL mismatch, SVA, and PT).

     This patient is classified as follows: L, ++, 0, +.
5. *What are the indications for and the goals of surgery?*
   - Indications include:
     - Persistent intractable back pain > 6 months with failed nonoperative measures
     - Persistent intractable radicular leg pain and/or neurogenic claudication symptoms (with concordant imaging findings)
     - Progressive deformity
     - Significant pulmonary dysfunction
   - Goals of surgery include:
     - Pain relief
     - Improvement in mobility, function status, and quality of life
     - Deformity correction with restoration of coronal and sagittal balance
     - Neural element decompression
6. *Describe the surgical options for deformity correction.*
   - Single thoracic curves (T)
     - Curve < 70 degrees: Can be managed with single stage posterior spinal fusion and instrumentation. Facet osteotomies are necessary to achieve an optimal correction.
     - Curve > 70 degrees: A combined anterior + posterior approach (single or two stage) may be required to obtain optimal correction. Vertebral column resection allows for sufficient mobilization to restore coronal balance.
   - Thoracolumbar and lumbar curves (L)
     - Either posterior/anterior/combined approach may be employed. The combined approaches are preferred for patients with poor bone quality and severe structural deformities (> 50 degrees).
   - Double thoracic and lumbar curves (D)
     - T/L curve < 60 degrees: Posterior spinal instrumentation and fusion is sufficient in the majority of cases.
     - T/L curve > 60 degrees: A combined anterior/posterior approach (single or two stage) may be required to obtain optimal correction. The first stage may involve anterior arthrodesis with structural allograft/cages and an internal fixation device. The second stage mainly involves posterior instrumentation and fusion.
   - Extent of fusion
     - To minimize the risk of proximal junctional kyphosis, the proximal extent of the instrumentation is not stopped at either the T/L junction or midthoracic apex area. The optimal proximal location of the upper instrumented vertebra is around either the lower thoracic or upper thoracic region.
   - Inclusion of sacrum is considered in the following cases:
     - L5–S1 degenerative disc disease
     - L5–S1 segmental instability
     - Uncompensated lumbosacral curvature
   - Inclusion of ilium
     - Inclusion of the ilium is considered a reasonable option to minimize the risk of implant failure and/or pseudoarthrosis in long segment constructs stopping at S1.
   - Surgery performed in this patient
     - T10 to iliac instrumented arthrodesis with multilevel laminectomies, facet (Ponte) osteotomies, and interbody cage placement at the L3–L4 and L4–L5 levels (▶**Fig. 118.4**)
7. *Describe the complications associated with adult spinal deformity (ASD) surgery.*
   - Early
     - Dural tear (0.4–15%)
     - Wound infections (1–16%)
     - Pulmonary (0.1–11%)
     - Renal (1–20%)
     - Neurological (1–11%)
     - Gastrointestinal (1–8%)
     - Cardiac (1–13%)
     - Mortality (< 1%)

## Answers (*continued*)

- Delayed
  - Pseudoarthrosis (5–27%)
  - Proximal junctional kyphosis (18.7–52.6%)
  - Proximal junctional failure (5.6–21%)
  - Distal junctional kyphosis (10.2%)

8. *What are the health-related quality of life (HRQOL) measures and instruments utilized to assess functional outcomes?*
   - The most commonly used HRQOL measures used to assess functional outcomes in ASD patients are as follows:
     - Oswestry Disability Index (ODI): The ODI questionnaire consists of eight spheres, i.e., pain intensity, personal care, walking/running, sitting, standing, sleeping, and travelling. Each item consists of six statements corresponding to scores from 0 to 5 which represent the least to maximum disability.
     - Scoliosis Research Society (SRS)-22: The SRS-22 questionnaire is the most commonly used tool to measure the overall impact of spinal deformity on a patient. The questionnaire includes five spheres (pain, function, self-image, mental health, and satisfaction) consisting of 22 questions. Scores of 1 to 5 represent a scale from maximum to least disability.
     - Short-Form Health Survey (SF-36): The SF-36 includes 36 questions in eight spheres: vitality, physical functioning, bodily pain, general health perceptions, physical role functioning, emotional role functioning, social role functioning, and mental health. All questions are scored on a scale from 0 to 100, with 100 representing the highest level of functioning.
9. *What are the radiological factors that best correlate with functional outcomes?*
   - According to Dubousset's theory of the "conus of economy," normal human posture assumes a stance limited within a narrow AP range to minimize muscle exertion. Thus, the restoration of various radiological parameters and realignment of the spinal axis are expected to influence clinical and functional outcomes in ASD patients. Radiological factors identified to date as having significant correlations with HRQOL outcomes include CSI < 4 cm, SVA < 5 cm, PI–LL mismatch = ±10 degrees, and PT < 20 degrees.

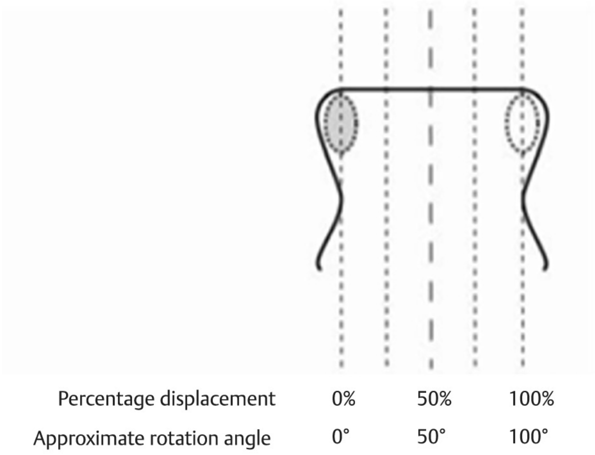

**Fig. 118.3** Nash–Moe method of radiological measurement of vertebral rotation.

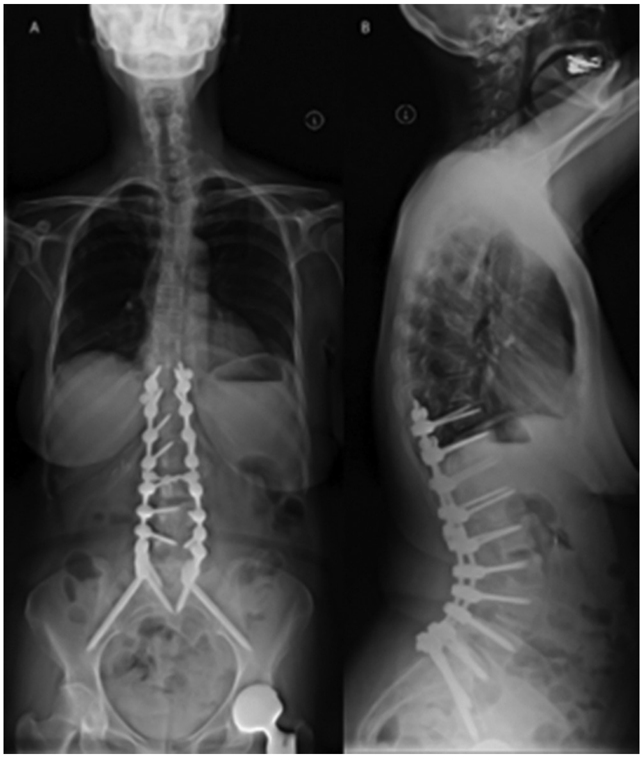

**Fig. 118.4** Postoperative full-spine standing anteroposterior and lateral X-rays showing T10–iliac instrumentation with correction of the CC angle and coronal spinal imbalance.

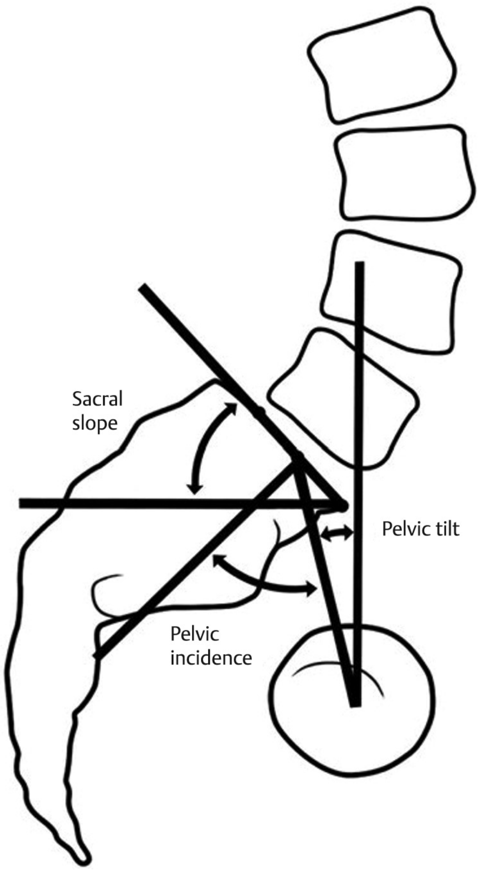

**Fig. 118.5** Rendering of Spinal pelvic parameters and their relationships. Pelvic Incidence = Pelvic Tilt + Sacral Slope. (Source: Spinal Pelvic Parameters. In: Benzel E, ed. Biomechanics of Spine Stabilization. 3rd Edition. Thieme; 2015.)

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# Case 119 Lower Back Pain—Conservative Management

*Hashem Al Hashemi, Remi Nader, and Abdulrahman J. Sabbagh*

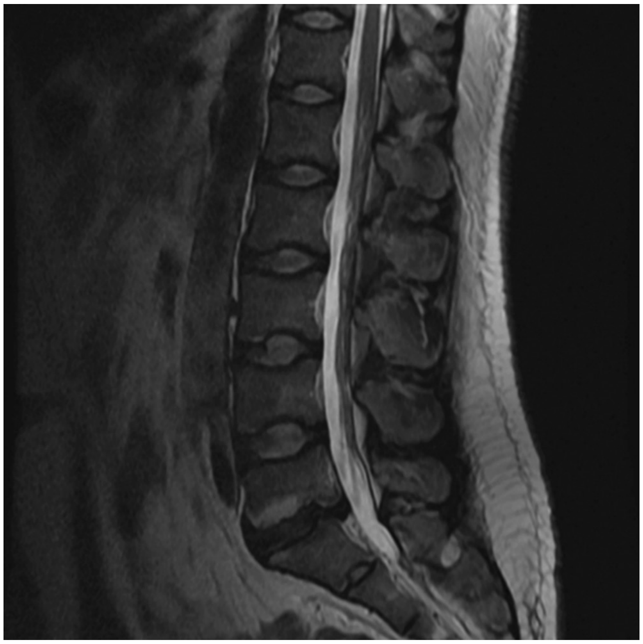

**Fig. 119.1** T2-weighted sagittal MRI of the lumbar spine demonstrating degenerative disc disease at the L5–S1 level.

## Clinical Presentation

- A 45-year-old man presents to your clinic complaining of lower back pain for the past 3 months.
- The patient's work activities do involve heavy lifting.
- He is otherwise neurologically intact.
- MRI of his lumbar spine was done prior to his visit; the image is shown in ▶**Fig. 119.1**.

## Questions

1. What are some common causes of lower back pain?
2. Provide a broad differential diagnosis for lower back pain.
3. What are some red flags or critical conditions that can present with lower back pain (on both history and physical examination)?
4. What imaging studies would you like to obtain?
5. What other investigations may be warranted?
6. Describe nonoperative measures of management of lower back pain.
7. What interventional procedures may be used to differentiate the types of spine-related lower back pain?
8. Discuss interventional options for management of spinal pain.

## Answers

1. *What are some common causes of lower back pain?*
   Common causes include[1]:
   - Poor posture
     - Poor sitting or standing posture
     - Sleeping position and/or pillow positioning
     - Bending forward too long
     - "Hiking" your shoulder to hold the phone receiver to your ear
   - Excess weight
     - Pregnancy
     - Obesity
   - Sudden or strenuous physical effort
     - Improper lifting
     - Accident, sports injury, or fall
     - Carrying a heavy purse, briefcase, or backpack
   - Stress and muscle tension
     - Lack of muscle tone
     - Deconditioning
2. *Provide a broad differential diagnosis for lower back pain.*
   Differential diagnosis includes[1]:
   - Congenital
     - Scoliosis, kyphosis
     - Isthmic spondylolisthesis
     - Spina bifida
     - Short pedicle syndrome
   - Infectious
     - Discitis, osteomyelitis, tuberculosis
     - Pyelonephritis, urinary tract infections, psoas abscess
     - Endometritis
   - Traumatic
     - Lumbosacral fracture or dislocation
     - Spondylolisthesis
     - Muscle strain or sprain
     - Traumatic disc injury, facet injury and ligamentous injury
     - Osteoporotic vertebral body fracture
   - Tumor
     - Metastatic disease to the spine
     - Primary bone tumors of the spine
     - Abdominopelvic retroperitoneal tumors
   - Environmental
     - Repetitive heavy lifting at work
     - Poor posture, obesity
     - Prolonged riding in car or truck
   - Neurogenic
     - Herniated disc, spinal stenosis
     - Hypertrophied ligaments
     - Neuropathic pain, failed back syndrome
   - Drugs
     - Antivirals, antibiotics
     - Chemotherapeutics
     - Coumadin
   - Inflammatory
     - Osteoarthritis, rheumatoid arthritis
     - Ankylosing spondylitis
     - Systemic lupus erythematosus
   - Vascular
     - Abdominal aortic aneurysm
     - Arteriovenous malformation of the spine
     - Hemangioma, hematoma
   - Gynecologic
     - Menstruating
     - Endometriosis
     - Pregnancy
     - Tumors
   - Acquired and other
3. *What are some red flags or critical conditions that can present with lower back pain (on both history and physical examination)?*
   - Clinical symptoms on history[2–4]
     - Age older than 50 years
     - Cancer: history of cancer, unexplained weight loss, pain at multiple sites
     - Pain worsening at night and not mechanical in character
     - Immunosuppression: human immunodeficiency virus (HIV), steroid use, transplant patient
     - Infection: fever, night sweats, back tenderness, limited range of motion
     - Trauma: history of significant trauma or minor trauma in osteoporotic patients
     - Intractability
     - Cauda equina syndrome: bladder or bowel dysfunction, saddle anesthesia, leg weakness or pain
     - Other neurologic symptoms
   - Clinical signs on physical examination[2–4]
     - Saddle anesthesia
     - Incontinence
     - Fever (> 38°C)
     - Urinary retention
     - Muscular weakness
     - Bony tenderness (vertebral)
     - Very limited range of spinal motion
4. *What imaging studies would you like to obtain?*
   - Basic imaging studies include plain X-rays of the lumbar and possibly of the thoracic spine.[5]
   - MRI of the lumbar spine[2,5]
     - Is not indicated for nonspecific low back pain
     - Many people without symptoms show abnormalities on radiographs and MRI. The chances of finding coincidental disc prolapse increases with age.
     - MRI should be reserved for patients with red flag conditions and those with neurologic symptoms and/or signs severe enough to consider surgery.
     - In the presence of bowel and bladder incontinence, diffuse weakness suggestive of multiple roots involvement or upper motor signs, an urgent whole-spine MRI is recommended.
   - Flexion and extension radiographs of the lumbar spine: recommended in cases of spondylolysis or

## Answers (*continued*)

spondylolisthesis (as there is potential spine instability).

- CT scan of the lumbar spine recommended in trauma cases to evaluate for fracture and alignment or in certain degenerative conditions to assess pars integrity, osteophytes, etc.[5]
- Other studies depend on the suspected pathologies.
  - Bone scan: recommended in suspected cases of malignancy (history of malignancy, history of weight loss, or night pain)
  - Abdominal X-ray and renal ultrasound (US): if kidney stones are suspected
  - Vascular studies (ankle brachial index, MR angiography [MRA] or angiography): if the patient gives a history of vascular claudication (smokers, diabetics, history of stenting or vascular bypass).

5. *What other investigations may be warranted?*
   - Investigative measures strongly depend on the detailed history and examination obtained from the patient. Certain important tests are mentioned below. This list is, however, by no means exhaustive.
   - With history suggestive of osteoporosis (steroid use, previous osteoporotic fractures, family history of osteoporosis, or osteopenia on radiograph), a bone densitometry is recommended.
   - If the clinical picture is suggestive of connective tissue disease (multiple joints or skin manifestations), a screening should be done with some laboratory tests at the least, such as: erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), rheumatoid factor (RF), and antinuclear antibodies (ANA).
   - If you suspect multiple myeloma, a skeletal survey, total proteins, and protein electrophoresis should be completed.
   - If the patient is diabetic, control of blood glucose should be checked with hemoglobin A1–C.
6. *Describe nonoperative measures of management of lower back pain.*
   - Activity modification: This may involve taking 2 to 3 days off work, avoiding activities that provoke pain, doing a different task at work with less stress on the spine; may result in changing jobs.[6,7]
   - Pain control: This is important initially with the activity modification to allow the patient to mobilize more and do some strengthening exercises. This could be done with the help of passive therapeutic modalities while in physical therapy sessions, such as heat, cold, ultrasound, and transcutaneous electrical nerve stimulation (TENS) machines.
   - Physical therapy programs include[8,9]:
     - During the first month of symptoms: low-impact aerobic exercise can minimize debility due to inactivity, such as walking, bicycling, or swimming.
     - Also, conditioning exercises for trunk muscles may be done such as exercises to strengthen back extensors and abdominal muscles (the McKenzie techniques comprise a well-known set of such exercises).
     - During the first 2 weeks, these exercises may aggravate symptoms. Therefore, the recommended exercise quotas are gradually escalated, resulting in better outcomes than having patients simply stop when pain occurs.
   - Educating the patient is also important in improving and hastening recovery.
     - Explanation of the condition to the patient in understandable terms.
     - Positive reassurance, in most cases, that the condition will almost certainly subside.
     - Teach the patient about proper posture, sleeping positions, lifting techniques, and weight loss techniques, if overweight.
   - If there is no improvement with such conservative techniques, then resort to medications initially and later interventional procedures may be considered.
   - Maintenance/recurrence prevention programs should also be set up.
     - After some control of the pain is achieved using the activity modification and medications (±) interventions, a core strengthening program should be initiated.
     - This involves lumbar stabilization exercise: strengthening and training muscles that surround the spine can help support and protect the spine.
     - A simple way to start this is walking and performing general aerobic exercises.
     - Exercise has a good general effect on the human body that includes increasing the pain threshold and building resistance to pain.
     - The patient also has to modify unhealthy lifestyle habits and poor body mechanics that may have initiated the pain.
   - Medical management includes[5,10]:
     - Nonsteroidal anti-inflammatory drugs (NSAIDS)
     - Acetaminophen
     - Other anti-inflammatories or nonnarcotic pain medications such as tramadol and combination of the above
     - For radicular pain that affects sleep, amitriptyline or other antidepressant-type pain medications may be used.
     - Alternatively, gabapentin or pregabalin can be employed. Both have no significant interactions with other medications, but need to be avoided in patients with significant renal dysfunction.
     - Strong narcotics may be used in the short term in cases of intractable pain. These include morphine derivatives such as MS Contin (Purdue Pharma, Stamford, CT), hydrocodone, hydromorphone, etc. Fentanyl patches are contraindicated for narcotics naive patients.

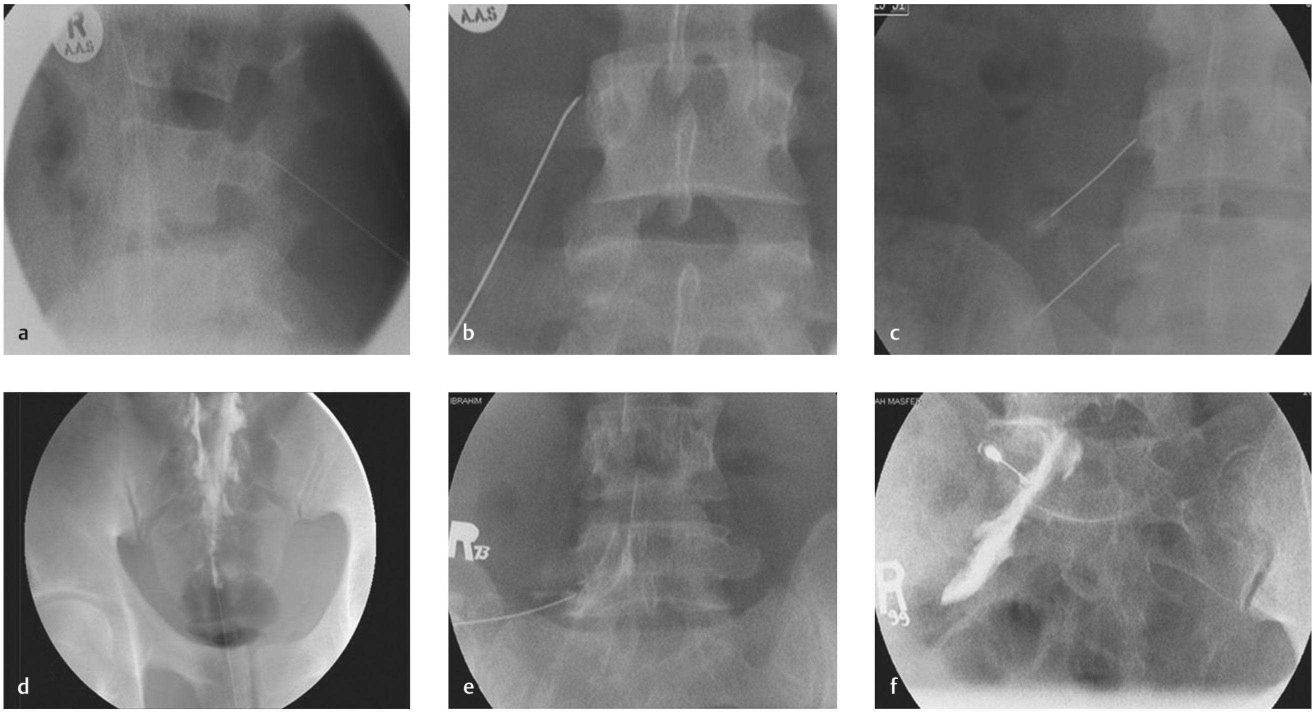

**Fig. 119.2** Plain radiographs taken while performing the following interventional procedures: (**a, b**) L3 medial branch neurotomy and (**c**) L3 and L4 medial branch blocks, (**d**) caudal epidural steroid injection, (**e**) L5 transforaminal epidural steroid injection, and (**f**) S1 transforaminal epidural steroid injection.

## Answers (*continued*)

7. *What interventional procedures may be used to differentiate the types of spine-related lower back pain?*
   - For mechanical back pain, the following measures may apply:
     – Medial branch blocks may help diagnose facet-related pain in cases of whiplash injuries and degenerative disc disease.
     – Sacroiliac (SI) joint injections may help diagnose and treat SI joint pain in cases of leg length discrepancy, multiple-level fusion, or spondylo-arthropathy (ankylosing spondylitis or inflammatory bowel disease).
     – Discography may be used to diagnose discogenic pain and to narrow the level of most severe involvement if surgical fusion is contemplated.
     – Trigger point injections may aid the diagnosis and treatment of muscular back pain.
8. *Discuss interventional options for management of spinal pain.*
   Options for interventional pain management include the following (▶**Fig. 119.2** for illustrative intraprocedural X-rays)[11,12,13]:
   - Transforaminal, caudal, or interlaminal epidural injections, for radicular back pain.[14,15]
     – Transforaminal and caudal epidural carries less risk for “wet” tap with spinal fluid leakage, spinal headache, and epidural hematomas.
     – Complications during epidural injections for back pain happen due to the following mistakes[16]:
       ○ Performing a blind epidural injection with no radiographic guidance
       ○ Not using contrast
       ○ Using particulate steroids such as methylprednisolone (Depo-Medrol; Pfizer Pharmaceuticals, New York, NY)
       ○ Other faulty techniques
     – There is no definite evidence that these procedures are effective in treating acute radiculopathy or lower back pain alone.
     – Epidural injections may be an option for short-term relief of radicular pain when control on oral medications is inadequate or for patients who are not surgical candidates.
   - Radiofrequency denervation (neurotomy) for facet related mechanical spinal pain that is proved by two positive medial branch blocks, using two different local agents of different duration.[17]
   - Spinal cord stimulators for treatment for chronic back pain[18]
   - Facet blocks to diagnose and treat facet related spinal pain
   - SI joint injection to diagnose and treat mechanical back pain that is related to sacroiliac joint disease
   - Vertebroplasy or kyphoplasty for management of osteoporotic compression spine fractures[19]
   - In general, interventional procedures are delayed for 4 to 12 weeks, depending on the case. It is essential to determine if the patient responds to more conservative measures first.
   - In proven intractable one-level discogenic pain, after failure of conservative measures, consideration could be made to anterior lumbar interbody fusion (ALIF) versus disc arthroplasty; the latter could only be considered in the absence of facet arthropathy.

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# Case 120 Lumbar Disc Herniation

*Patrick Kim, Ali Luqman, Jorge E. Alvernia, and Gustavo Luzardo*

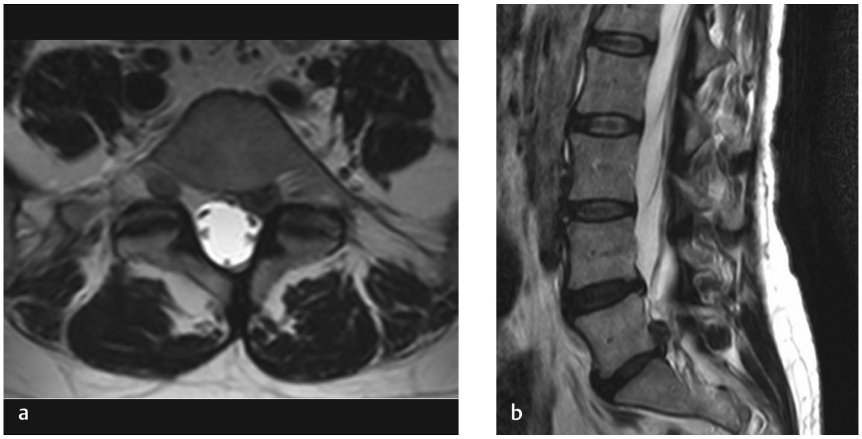

**Fig. 120.1** T2-weighted MRI axial (**a**) and sagittal (**b**) of patient's pathology. Depicted is a lumbar disc herniation at L5–S1 and a disc bulge at L4–L5. Note the change in disc intensity at these levels indicative of degenerative changes of these discs.

## Clinical Presentation

- The patient is a 50-year-old female who complains of 10/10 left-sided buttock pain that radiates down to the feet. This has been present since a fall 6 months ago. She also reports paresthesia down the back of her left leg and on the top of the foot.
- She has not had any symptom relief on nonsteroidal anti-inflammatory drugs (NSAIDs), steroids, or gabapentin. The patient attended physical therapy for 6 months but was unable to complete the exercises due to severe pain.
- An MRI is obtained and shown in ▸**Fig. 120.1.**

## Questions

1. How do symptomatic lumbar disc herniations (LDH) present? What are some of the symptoms and signs on history and physical examination? Which imaging modality is best suited to evaluate LDH?
2. How would you classify LDH? What type of LDH does this patient have?
3. What is the most common level(s) of LDH and into which zones?
4. What are the five zones of LDH? Into which zone does this patient's herniation extend?
5. What is the incidence of far lateral LDH? Why are these cases different?
6. What are the borders of the lateral recess? What is lateral recess syndrome? What measurements define lateral recess syndrome?
7. What is the initial management of symptomatic LDH?
8. Which cases necessitate urgent or emergent surgical intervention?
9. What are some surgical options for symptomatic LDH?
10. What are the most common complications of LDH surgery and their management?

## Answers

1. *How do symptomatic lumbar disc herniations (LDH) present? What are some of the symptoms and signs on history and physical examination? Which imaging modality is best suited to evaluate LDH?*
   - Most LDH present with radiculopathy from nerve root compression. The patient may complain of a constant, sharp, "electric" type feeling radiating from the buttocks down the leg in various patterns. Depending on whether the exiting and/or traversing nerve root is compressed, one of five lumbar disc syndromes may be described by the patient (▶**Table 120.1**). Patients may also present with back pain of varying degree.
   - There are a multitude of physical findings on clinical exam that may be present. Most, on their own, are unreliable and have poor sensitivity and/or specificity,[1–3] and correlation with imaging is paramount.
     - Lasegue's sign (ipsilateral straight leg raise): With the patient supine, the affected lower extremity (LE) is raised (flexing only the hip) until radicular pain is reproduced. Has high sensitivity, but low specificity. Pain is usually elicited at 60 degrees of elevation. Best utilized to evaluate for L5 and S1 radiculopathy.
     - Fajersztajn's sign (contralateral straight LE raise): With the patient supine, the contralateral leg is elevated in a similar fashion. Pain again is felt in the ipsilateral leg, but usually at a greater angle of about 90 degrees. Little sensitivity, but high specificity.
     - Reverse straight leg raise: This test is best utilized to assess upper LDH (L2–L4). In a prone patient, the ipsilateral LE is extended at the hip until radicular pain is reproduced. Can be fairly sensitive for detecting LDH even in the presence of a negative Lasegue's sign.
     - Decreased ankle/patellar reflex: Asymmetric reflexes or loss of a reflex can be useful in conjunction with other findings, but alone, is an unreliable sign.
     - Sensory deficit: Similar to the reflex examination, a sensory deficit alone can be a poor localizing sign, but may be useful when taken together with other findings.
     - Motor deficit: A focal motor deficit may be noted on examination. Although this is not a sensitive finding for localizing LDH, it does have a high specificity.
   - MRI is the gold standard for visualizing LDH. If MRI is contraindicated, a CT myelogram may be performed which can demonstrate a filling defect at the site of the disc herniation.
2. *How would you classify LDH? What type of LDH does this patient have?*
   - This patient has a left L5–S1 focal disc extrusion without sequestration.
   - There are two types of disc herniation, each with subclassifications:
     - Intervertebral disc herniation:
       - Focal: herniation of disc involving < 25% of disc circumference.
       - Broad-based: herniation of disc involving 25 to 50% of disc circumference.
       - These can be further classified as:
         - Protrusion: a disc herniation that has no neck or focal narrowing.
         - Extrusion: a disc herniation with a narrow segment or neck; these may also be *sequestered* (a free fragment, or disconnection from originating disc space) and/or *migrated*.
     - Intravertebral disc herniation: also known as a Schmorl's node, this is a herniation of disc material in a cephalad or caudal direction into the adjacent endplates. These are very common, and largely asymptomatic, considered indicative of lumbar disc disease/degeneration.
3. *What is the most common level(s) of LDH and into which zones?*
   - Most LDH occur at the L5–S1 and L4–L5 levels (> 90%), and into the posterolateral zones (paramedian and foraminal) (> 90%). Upper lumbar discs represent a relatively small minority of LDH.[4,5] Extreme lateral/far lateral discs comprise about 10% of LDH.[6] Symptomatic central LDH are also relatively rare comprising 1.2% of surgical LDH.[7]
4. *What are the five zones of LDH? Into which zone does this patient's herniation extend?*
   - This is a case of posterolateral LDH involving both the paramedian and foraminal zone.
   - There are five zones of disc herniation (▶**Fig. 120.2**) and they affect either the traversing or exiting nerve roots.
     - Central (canal): affects traversing nerve root(s) (level below disc space, preganglionic)
     - Paramedian (lateral recess/subarticular): affects traversing nerve root (level below, preganglionic)
     - Foraminal: compression of exiting nerve root (same level, preganglionic)
     - Far lateral (extraforaminal/extreme lateral): compression of exiting nerve root (same level, may also compress dorsal root ganglion)
     - Anterior: no nerve root compression, may affect surrounding tissues and paraspinal musculature.
5. *What is the incidence of far lateral LDH? Why are these cases different?*
   - Far lateral or extreme lateral LDH can account for up to 10% of all disc herniations.[6] The exiting nerve root is affected in the extraforaminal zone and there can also be compression of the dorsal

**Table 120.1** Lumbar disc syndromes based on affected nerve root and associated clinical findings

| Nerve Root | Incidence[4,5] | Reflex Involved | Motor Deficit | Sensory/Pain Distribution |
|---|---|---|---|---|
| L2 | < 1% | Cremaster | Hip flexion | Anterior/medial upper thigh, lateral groin |
| L3 | < 1% | Patellar (knee-jerk) | Hip flexion, lateral rotation of hip | Anterior thigh above knee, lower medial thigh |
| L4 | 3–8% | Patellar (knee-jerk) | Knee extension | Anterior/lateral thigh, anterior leg, medial ankle/malleolus and foot |
| L5 | 35–47% | Medial hamstring | Dorsiflexion, extensor hallucis longus | Lateral/posterior thigh, lateral leg, dorsum of foot and large toe |
| S1 | 47–58% | Ankle jerk (achilles) | Plantar flexion | Posterior thigh, posterior leg, lateral ankle/malleolus and foot |

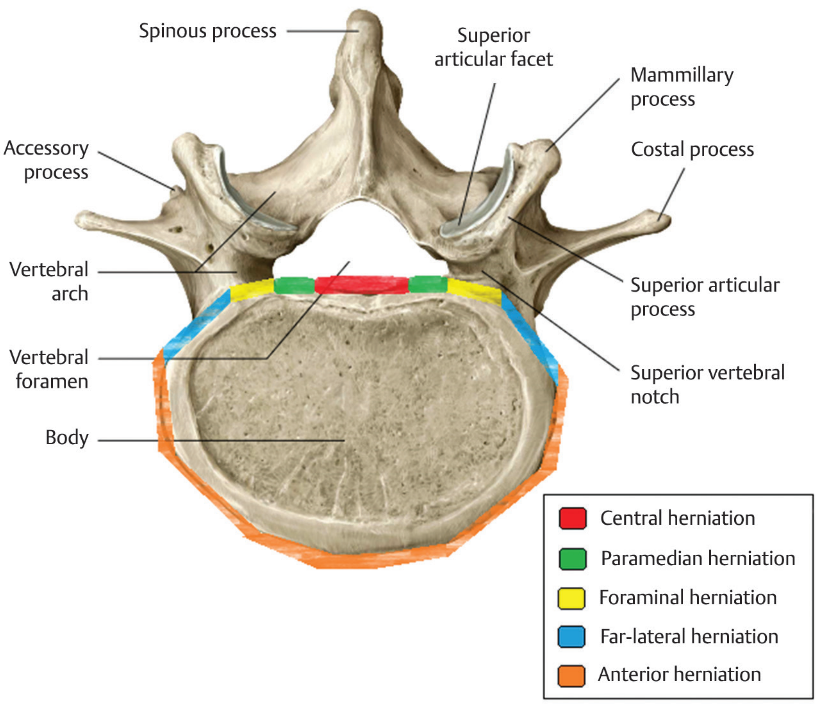
Spinous process
Superior articular facet
Mammillary process
Costal process
Accessory process
Vertebral arch
Superior articular process
Vertebral foramen
Superior vertebral notch
Body
🟥 Central herniation
🟩 Paramedian herniation
🟨 Foraminal herniation
🟦 Far-lateral herniation
🟧 Anterior herniation

**Fig. 120.2** Axial anatomical image of typical lumbar vertebrae and the zones of herniation.

## Answers (*continued*)

root ganglion, producing more severe pain on presentation.

- LDH in this zone are more likely to affect the upper lumbar nerve roots (L2 and L3 account for 32% of these cases[6]), although L4 remains the most commonly affected nerve root (60%). Lasegue's sign is usually negative, but pain can be reproduced with lateral bending.
- These types of herniations have a higher incidence of double herniation. They pose a unique diagnostic and treatment situation, usually requiring lateral approaches.

6. *What are the borders of the lateral recess? What is lateral recess syndrome? What measurements define lateral recess syndrome?*
   - Lateral recess: bordered anteriorly by the vertebral body, laterally by the pedicle, and posteriorly by the superior articular facet of the inferior vertebral body; defined as the area just proximal to where the nerve root exits through the neural foramen.
   - Lateral recess syndrome develops when hypertrophy of the superior articular facet causes compression of the nerve root. This results, most commonly, in leg pain predominantly when walking or standing which is relieved by sitting and flexion.
   - To measure the dimension of the lateral recess, measure the anterior border (vertebral body) to posterior (superior articular facet) border distance using the bone window on CT. Lateral recess syndrome is diagnostic at a measurement < 2 mm, and is suggested at < 3 mm, and a dimension of 3 to 4 mm is considered borderline.[8]

## Answers (*continued*)

7. *What is the initial management of symptomatic LDH?*
   - Without any "red flag" symptoms, no further testing is recommended. Red flags include, but are not limited to:
     - Acute-onset urinary retention or overflow incontinence
     - Fecal incontinence
     - Reduction or loss of anal sphincter tone
     - Saddle anesthesia
     - Sexual dysfunction
     - Progressive weakness in the lower extremities
     - Any history of trauma, cancer, or infection
   - In the absence of "red flags," initial management of LDH is conservative, and should be initially implemented for at least 4 weeks (90% of cases will resolve spontaneously).[1] Conservative treatment may include:
     - Acutely, bed rest for no more than 2 to 3 days
     - Physical therapy which may include: exercise, education, manipulation, aqua therapy, massage
     - Activity modification: posture training, limitations on heavy lifting, bending and twisting, and avoidance of prolonged sitting
     - Medications: analgesics (acetaminophen or NSAIDs), muscle relaxants, neuropathic pain medication (gabapentin, Lyrica). Narcotic use should be avoided.
     - Interventional pain management: epidural steroid injections, nerve root blocks (diagnostic and therapeutic)
8. *Which cases necessitate urgent or emergent surgical intervention?*
   - Cauda equina syndrome is a neurosurgical emergency. It can result from a herniated disc: L4–L5 (59%), L5–S1 (30%),[9–11] spinal canal stenosis or foraminal stenosis (very rarely). Early surgery is recommended but not absolute (no clear correlation between time to intervention and recovery).[9] The generally accepted management in acute cauda equina is intervention within 24 to 48 hours after syndrome onset.[9,10,11]
   - Acute and profound or progressive neurologic deficit (in particular, a motor deficit) may necessitate urgent surgical intervention.
9. *What are some surgical options for symptomatic LDH?*
   - For LDH without spinal stenosis:
     - Standard discectomy or microdiscectomy (similar efficacy[12])
   - For foraminal or far lateral LDH:
     - Partial or total facetectomy
     - Lateral (extracanal) approach
   - For LDH and spinal stenosis
     - Decompressive laminectomy with or without discectomy
     - Fusion may be indicated for stenosis with associated instability
10. *What are the most common complications of LDH surgery and their management?*
    - Intraoperative complications
      - Wrong level surgery, durotomy, hemorrhage, nerve root, vessel and/or visceral injuries, death (rare)[13]
    - Postoperative complications
      - Immediate: nerve root damage, complications from positioning, bladder function disturbance, cauda equina syndrome, thromboembolism (0.1–1%), wound infection, epidural hematoma, abdominal content injury.[14]
      - Late: thromboembolism, recurrent disc, failed back syndrome (postdiscotomy syndrome), meningocele due to unrecognized cerebrospinal fluid (CSF) leak, macroinstability, iatrogenic instability.

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# Case 121 Black Disc with Advanced Modic Changes

*Rory Mayer and Ibrahim Omeis*

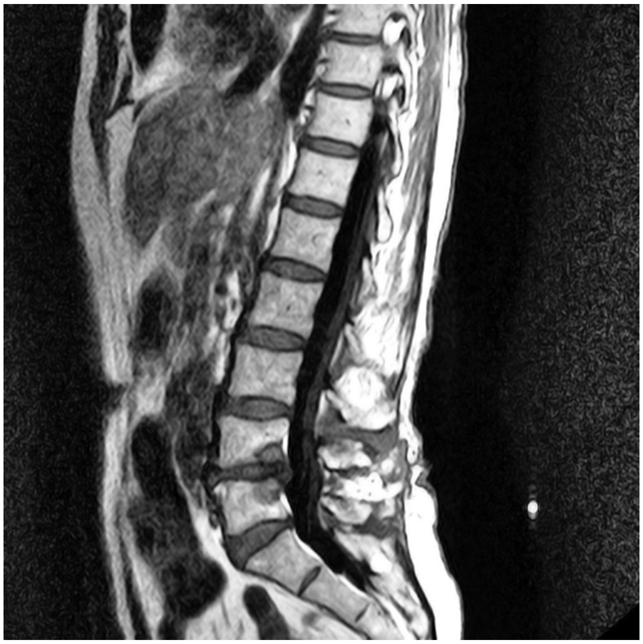

**Fig. 121.1** MRI T1-weighted image, sagittal view of the lumbar spine.

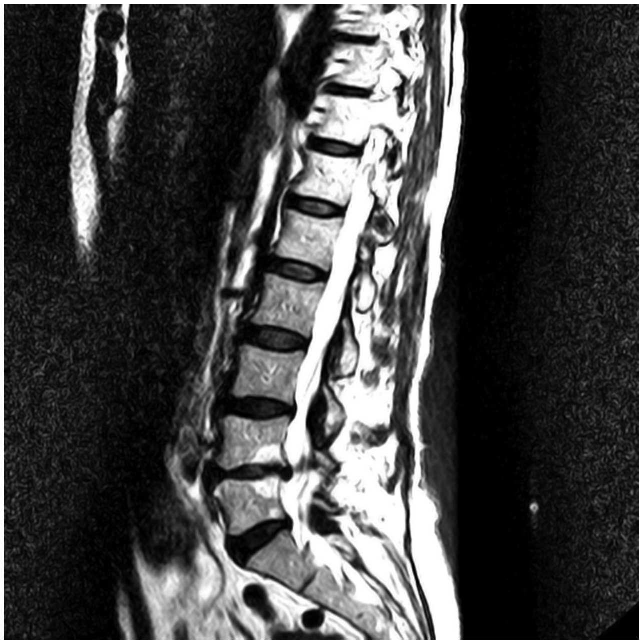

**Fig. 121.2** MRI T2-weighted image, sagittal view of the lumbar spine.

## Clinical Presentation

- A 56-year-old female is referred to clinic with severe back pain refractory to conservative measures including physical therapy and medication.
- The patient describes back pain that is not associated with leg pain or numbness.
- She reports a worsening of her symptoms while bending forward and with running. Her pain improves minimally when recumbent.
- Her physical exam, including neurological exam, is unremarkable. Notably, there is no tenderness to palpation in the paraspinal lumbar region.
- An MRI scan is obtained and shown in ▶ **Fig. 121.1.**

## Questions

1. Describe the MRI (▶ **Fig. 121.1**).
2. Define the Modic grading system used to describe degenerative disc disease.
3. What is the appropriate classification of this patient's MRI?
4. Briefly describe the pathophysiology of the "black" disc.
5. What are the important findings from history and physical exam for patients with discogenic back pain?
6. How will you work up this patient, including any additional tests you may like?
7. What conditions are important to rule out while evaluating this patient?
8. What treatment options will you offer this patient with intractable back pain and advanced Modic changes with a black disc on MRI?
9. Why is it important to fully evaluate the patient's psychological and social situation prior to committing to treatment?
10. What are the appropriate considerations in counseling an asymptomatic patient with advanced Modic changes on MRI?

## Answers

1. *Describe the MRI* (▶**Fig. 121.1**).
   - The MRI demonstrates a T1- and T2-weighted images (T1WI and T2WI) of the lumbar spine in the sagittal view. There are hypointensities on T1WI (▶**Fig. 121.1**) and hyperintensities on T2WI (▶**Fig. 121.2**) at the L4–L5 vertebral bodies as well as hypointense or black L4–L5 disc. On this view, there is no evidence of intervertebral disc herniation, foraminal stenosis, or significant degenerative changes at other levels, or canal stenosis. Collectively, these findings are consistent with advanced Modic changes and degenerative disc disease isolated to the L4–L5 level.
2. *Define the Modic grading system used to describe degenerative disc disease.*
   Degenerative disc disease can be described using a three-stage system proposed by Modic and colleagues.
   - Type I disease demonstrates abnormal marrow changes with hypointensity on T1WI and hyperintense signal changes on T2WI that are consistent with edema and inflammation.
   - Type II disease demonstrates hyperintense signal on T1 and isointense or hyperintense signal on T2WI and reflects reactive fatty deposition in the marrow.
   - Type III disease describes advanced degenerative change at the intervertebral level with hypointense signal on both T1- and T2-weighted sequences consistent with sclerosis (see ▶**Fig. 121.3**).[1–3]
3. *What is the appropriate classification of this patient's MRI?*
   The MRI is consistent with Modic type I change due to the hypointense marrow sequence on T1WI and hyperintense signal on the T2WI and represents advanced degenerative disease.
4. *Briefly describe the pathophysiology of the "black" disc.*
   - The "black" disc represents the radiographic finding of decreased T2 signal that may result from degeneration due to any of the following causes: disc desiccation, disc space narrowing, calcification, endplate sclerosis, and annular degeneration or tears.
   - Cytokines and nitric oxide may play a role in the pain response, and changes in proteoglycan content with water loss and decreased collagen strength which precipitate disc degeneration.[4]
5. *What are the important findings from history and physical exam for patients with discogenic back pain?*
   - The history of discogenic back pain is characterized by either an acute event involving axial loading followed by intractable back pain or chronic progressive symptoms that worsen with sitting but improve with recumbency.
   - Activities that may exacerbate the pain include flexion or twisting of the lumbar spine, coughing, running, or the climbing of stairs. All of these activities increase axial load on the intervertebral disc and intradiscal pressure.
   - Pertinent physical exam findings in these patients are limited; a decreased range of motion in the lumbar region may be evident and although rare, paraspinal tenderness to palpation may occur. Annular tears may result in pain radiating down the dorsal thighs.[5]
6. *How will you work up this patient, including any additional tests you may like?*
   - Standing radiographs with flexion-extension views should be included to evaluate sagittal and coronal plane alignment as well as listhesis.
   - In the absence of any other pathology on imaging, the next step may include provocative testing with discography. Discography involves the injection of dye into the intervertebral disc while simultaneously correlating injection with the inducement of pain. It has been shown to be useful to differentiate a concordant pain-generating disc from nonconcordant healthy discs; however, the test can be equivocal. Recent evidence suggests some increased risk of accelerated disc degeneration after discography and patients should be counseled regarding this concern.
   - Post-discography CT scanning to evaluate for dye extravasation consistent with annular tears may also be a useful adjunct to guide decision-making.[4,6]
7. *What conditions are important to rule out while evaluating this patient?*
   - It is important to rule out musculoskeletal pain related to myofascial or ligamentous etiology; these patients demonstrate tenderness on exam.
   - Facet arthropathy will exhibit worsening of pain with extension maneuvers. Isthmic spondylolisthesis may manifest with acute or chronic back pain with or without radiculopathy.
   - A black disc and Modic change may represent an early sign of global sagittal or coronal malalignment; therefore, plain radiographs should be reviewed to rule out any overt signs of deformity.[7]
8. *What treatment options will you offer this patient with intractable back pain and advanced Modic changes with a black disc on MRI?*
   - Nonoperative treatment options include methylene blue injection, steroid injection, ramus communicans ablation, intradiscal electrothermal therapy, and biacuplasty; however, although some studies favor intervention, there remains no clear data as to whether these treatments confer stable long-term benefit. These options are best suited for younger patients with minimal loss of disc height.[5,8]

## Answers (*continued*)

- Surgical considerations include open or minimally invasive fusion at the pathologic level with removal of the disc and replacement with a graft. An interbody graft may be placed via an anterior, lateral, transformational, or posterior approach; however, there is limited data on superiority of one technique over another for discogenic disease. If there is concern for facet arthropathy as an additional pain source or the surgeon seeks immediate stability for mobilization, the use of posterolateral screw fixation to create a circumferential fusion is appropriate. Motion-sparing treatment with lumbar disc arthroplasty is an alternative option.[6,9–11]

9. *Why is it important to fully evaluate the patient's psychological and social situation prior to committing to treatment?*
   - Multiple studies have demonstrated that psychosocial factors can significantly impact a patient's perception of pain. Therefore, it is appropriate to screen for depression and fibromyalgia. In addition, these patients, if untreated, may have poor responses to surgery.
   - Screening should also identify patients' ongoing litigation related to employment or disability as these findings can be at times correlated with poor surgical outcomes.[4,6]
10. *What are the appropriate considerations in counseling an asymptomatic patient with advanced Modic changes on MRI?*
    - Studies have demonstrated that MRI findings including a black disc or Modic changes in an asymptomatic patient do not correlate with later symptomatic lumbar spine disease.
    - Therefore, patients with these findings should be followed up and further work-up be delayed until they become symptomatic.[7]

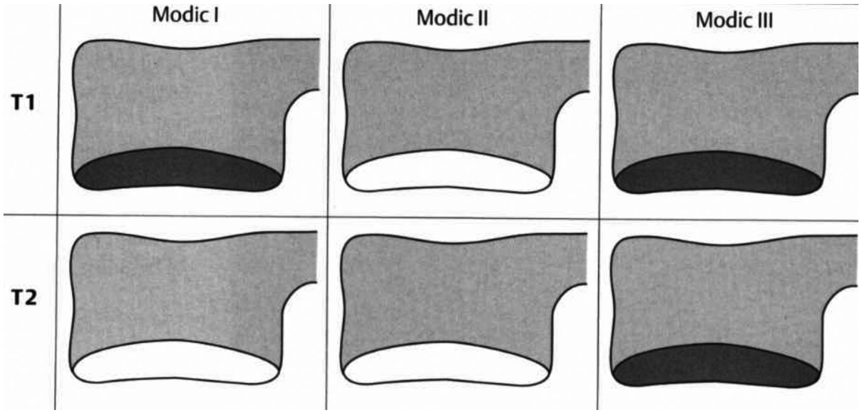

**Fig. 121.3** Illustration of Modic changes on MRI. Modic 1: edema. Modic 2: fatty marrow conversion. Modic 3: sclerosis. (Source: Imaging Signs. In: Imhof H, ed. Direct Diagnosis in Radiology. Spinal Imaging.. 1st Edition. Thieme; 2007.)

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# Case 122 Neurogenic versus Vascular Claudications

*Eric P. Roger and Edward C. Benzel*

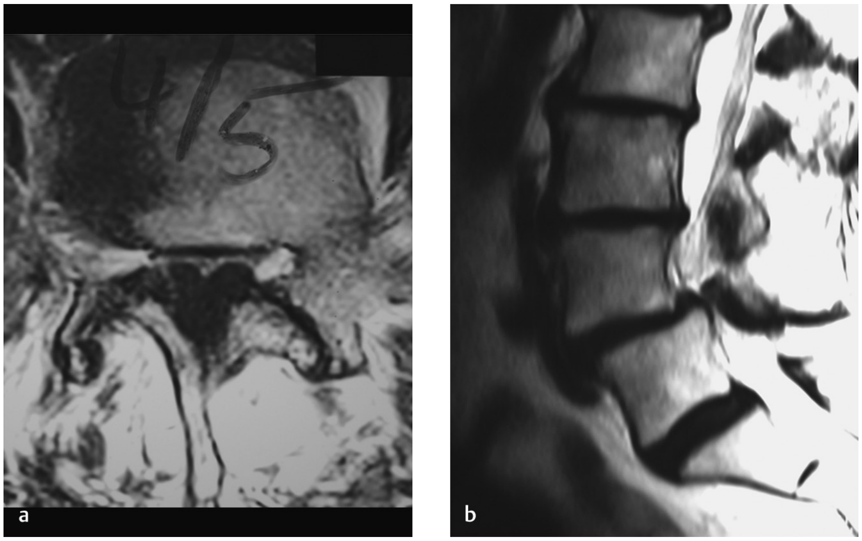

**Fig. 122.1** (**a**) Axial T2-weighted MRI at the L4–L5 disc space. (**b**) Sagittal T2-weighted MRI of the lumbar spine at the level of the midline.

## Clinical Presentation

- A 69-year-old man presents with difficulty ambulating.
- The patient also complains of leg "tightness" when he stands or walks, which completely resolves with sitting or leaning over a countertop.
- He has no back pain and no bladder or bowel dysfunction.
- An MRI scan is obtained and shown in ▶**Fig. 122.1**.

## Questions

1. Interpret the MRI.
2. What other radiologic imaging would you order?
3. What is the differential diagnosis of his leg symptoms?
4. What elements of the clinical presentation would help differentiate spinal versus vascular type of claudication?
5. What other diagnostic modality would help differentiate the two?
   The patient has no evidence of arterial insufficiency on examination and has a positive "shopping cart" sign (resolution of the leg pain when leaning on a shopping cart while walking). He is able to use a stationary bicycle without pain or symptoms. His ankle brachial indices (ABIs) are 1.10 on the right, 1.08 on the left side.
6. What therapeutic options are available to this patient?
7. What is the likelihood of destabilizing the spondylolisthesis with a simple decompressive laminectomy? What are some risk factors of this procedure?
8. Should a fusion be added to his surgical treatment? Should instrumentation also be added?

## Answers

1. *Interpret the MRI.*
   - The images demonstrate a grade 1 spondylolisthesis of L4 over L5.
   - There is no element of pars defect, at least on these images.
   - There is resulting severe central stenosis at that level, and to a lesser extent at the level above (L3–L4).
   - Paracentral cuts would be required to fully assess the amount of foraminal stenosis present.
2. *What other radiologic imaging would you order?*
   - Plain radiographs would be essential in the evaluation of this patient. Standing anteroposterior, lateral, and oblique views should be obtained.
   - In addition, dynamic films (flexion and extension views) would be useful.
   - MRI may underestimate the degree of listhesis as the images are performed in the supine position. Standing radiographs may show an accentuation of the L4–L5 slip, therefore suggesting instability.
   - This may be confirmed by flexion-extension (flex-xt) views.
   - Oblique views may be useful to assess a pars defect with a lysis seen across the neck of the "Scotty dog" appearance of the vertebrae.
   - CT scan of the lumbar spine can also be used to assess the pars integrity.
3. *What is the differential diagnosis of his leg symptoms?*
   - Although the differential diagnosis of leg "tightening" and similar symptoms can be attributed to a vast array of etiologies, including neurodegenerative disorders, hip and/or knee pathology, metabolic causes, etc., the two most likely diagnoses are neurogenic and vascular claudications.[1]
4. *What elements of the clinical presentation would help differentiate spinal versus vascular type of claudication?*
   - Vascular claudications[1]
     - Symptoms
       - Calf pain while walking (classic)
       - Buttock, thigh, and/or foot pain may also be present.
       - Brought on by walking or other activity
       - Relieved within a few minutes of rest
       - Numbness and/or weakness may be present.
       - Rest pain in advanced cases
     - Signs
       - Decreased pulses and bruits
       - Skin changes of arterial insufficiency (shiny, glistening, atrophic, ulcerated, loss of hair, nail changes)
       - Pallor when elevated
       - Dusky rubor when placed in a dependent state (Buerger's sign)
   - Neurogenic claudication[1]
     - Symptoms
       - Leg pain while standing or walking
       - May be described as weakness or "giving out"
       - Often associated with numbness, potentially dermatomal if stenosis is foraminal
       - Brought on by walking and/or standing
       - Relieved relatively quickly with sitting or bending forward
     - Signs
       - Often normal on examination
       - May have diminished deep tendon reflexes
       - May have dermatomal paresthesias
       - May have positive straight leg raising (Lasègue's sign)
       - Wide-based gait
   - Differentiation of neurogenic versus vascular claudications[2]
     - Symptoms
       - Neurogenic aggravated by standing or walking; vascular aggravated by walking but not just standing.
       - Neurogenic improves faster after sitting than does vascular.
       - Neurogenic improves by walking bent forward, for example, with the use of a shopping cart ("shopping cart sign").
       - Vascular aggravated by use of stationary bicycle; not neurogenic
       - Vascular claudication patients "look vasculopathic," with skin changes, nail changes, hair loss. Diminished pulses may be noted.
       - Neurogenic claudication patients often have a relatively normal examination, although neural deficits may be noted.
5. *What other diagnostic modality would help differentiate the two?*
   - ABI: ratio of Doppler-measured blood flow in upper versus lower extremities
     - Normal ratio is > 1.
     - A ratio of 0.6 to 0.9 indicates the claudicant range.
     - A ratio ≤ 0.5 correlates with rest pain and ulceration.
6. *What therapeutic options are available to this patient?*
   - Conservative management[3]
     - Medications (unlikely to be beneficial): include nonsteroidal anti-inflammatory drugs, gabapentin, other antiepileptics use to treat neuropathic pain, or other pain medications.
     - Therapies[4,5]
       - Physical therapy
       - Pilates therapy
       - Exercise therapy
       - Chiropractic therapy
       - Traction
     - Injections[5]
       - Epidural
       - Selective nerve root blocks

## Answers (*continued*)

- Surgical management[3,6]
  - Decompression (open or minimally invasive)[7,8,9]
    - Laminectomies[10]
    - Laminotomies and foraminotomies[2]
    - Laminoplasty
    - Interspinous decompression via a spinous process spacer device[11]
  - Decompression and fusion[12]
- Noninstrumented
  - Facet fusion
  - Posterolateral onlay fusion
- Instrumented[13,14]
  - Posterolateral fusion (PLF)
  - Posterior lumbar interbody fusion[13]
  - Transforaminal lumbar interbody fusion (TLIF)
  - Anterior lumbar interbody fusion (indirect decompression), not generally recommended in these cases

7. ***What is the likelihood of destabilizing the spondylolisthesis with a simple decompressive laminectomy? What are some risk factors of this procedure?***
   - The likelihood of destabilizing a low-grade (I or II) spondylolisthesis after laminectomy is poorly reported in the literature.
   - There are certain factors that may predispose to an increased risk of destabilization.[15]
     - Preoperative:
       - Pars defect (isthmic)
       - Evidence of instability on flexion-extension films: > 4.5 mm of translation[16]
       - Presence of a tall disc space
       - Sagittally oriented facets
       - Overweight patient
     - Intraoperative:
       - Partial or generous facetectomy
       - Injury to the pars interarticularis
       - Overt evidence of instability

8. ***Should a fusion be added to his surgical treatment? Should instrumentation also be added?***
   - Although the rate of spondylolisthesis progression after simple decompression is unclear, several studies have reported improved clinical outcomes after fusion (with or without instrumentation).
   - The Lumbar Fusion Guidelines[17] have therefore stated the following:
     - "Guidelines: The performance of a lumbar PLF is recommended for patients with lumbar stenosis and associated degenerative spondylolisthesis who require decompression. There is insufficient evidence to recommend a treatment guideline."[17]
   - It would appear that the addition of instrumentation to the fusion procedure increases the radiologic rate of fusion, although most studies have shown no statistical improvement in clinical outcome.
   - In this regard, the Lumbar Fusion Guidelines[17] have stated the following:
     - "Options. Pedicle screw fixation as an adjunct to lumbar PLF should be considered as a treatment option in patients with lumbar stenosis and spondylolisthesis in cases in which there is preoperative evidence of spinal instability or kyphosis at the level of the spondylolisthesis or when iatrogenic instability is anticipated."[17]
   - A prospective randomized control trial on 66 patients, the Spinal Laminectomy Versus Instrumented Pedicle Screw Fusion Study, has stated the following conclusions: in patients with degenerative grade I spondylolisthesis, the addition of lumbar spinal fusion to laminectomy was associated with a slightly better but clinically meaningful improvement in overall physical health-related quality of life when compared to laminectomy alone.[18]

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# Case 123 Cauda Equina Syndrome

Cristian Gragnaniello, Anthony M. T. Chau, Mohammad Almubaslat, and Remi Nader

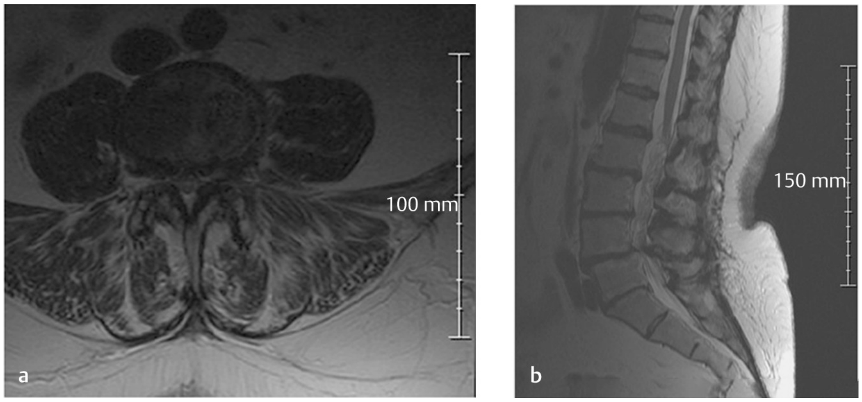

**Fig. 123.1** (**a**) Axial T2-weighted MRI at the L5 pedicle level. (**b**) Sagittal T2-weighted MRI through the lumbar spine.

## Clinical Presentation

- A 63-year-old previously well woman presents with acute-onset bilateral L5 radiculopathy and difficulty voiding after heavy lifting while gardening.
- On examination, there was mild weakness of bilateral great toe dorsiflexion and pinprick perineal paresthesia. A postvoid residual ultrasound after 300 mL of urinary output demonstrated 500 mL retained in the bladder.

## Questions

1. What is the clinical diagnosis? Describe the main clinical features of this diagnosis.
2. What conditions can cause these symptoms and signs?
3. What imaging is required?
4. Interpret the MRI findings (▶**Fig. 123.1**).
5. How would you proceed?
6. How would you counsel the patient regarding her prognosis?
7. Describe bladder micturition physiologic mechanisms.

## Answers

1. *What is the clinical diagnosis? Describe the main clinical features of this diagnosis.*
   - Acute cauda equina syndrome (CES) with urinary retention.
   - The syndrome consists of various combinations of back pain, uni- or bilateral radiculopathy, bladder or bowel disturbance, sexual dysfunction, and saddle region paresthesia. Generally, micturition dysfunction is required.
   - CES can be divided into two stages: incomplete CES (CESI) and CES with retention (CESR). CESI patients describe altered urinary sensation or straining micturition, while CESR patients describe the end-state syndrome, with painless urinary retention, overflow incontinence, and complete loss of bladder control.
2. *What conditions can cause these symptoms and signs?*
   - CES may present in three ways: suddenly without prior complaints, in patients with chronic low back problems, and either acute or insidious bladder dysfunction.
   - The etiology is frequently compressive (▶**Table. 123.1**). The most common cause especially in acute CES is a disc herniation; 90% of the time it will be at L4–L5 or L5–S1. About 2% of operated herniated discs are for CES.

## Answers (*continued*)

3. *What imaging is required?*
    - Even though audits document a 90% negative result for emergency MRIs in cases of suspected CES, an emergency MRI is required in acute CES even in the middle of the night.
    - This patient exhibits both saddle region paresthesia and urinary retention of 500mL—both strong positive predictors of CES. There are no reliable negative predictors.[1]
4. *Interpret the MRI findings (▶* **Fig. 123.1***).*
    - Grade 1 degenerative spondylolisthesis at L4–L5 causing severe narrowing of the canal and nerve root impingement.
    - The nerve root impingement correlates very well with the clinical signs of radiculopathy.
    - A sign that the spondylolisthesis is due to degenerative changes can be found in the sagittal image that shows a malalignment of the spinous process of L4 that displaces with the vertebral body due to maintenance of the integrity of the neural arch.
5. *How would you proceed?*
    - Emergency decompressive L4–L5 interlaminar decompression and discectomy
    - Recent consensus is that especially in cases of acute urinary dysfunction, emergency surgery is indicated. Previously cited literature stating a blanket 48-hour safety window for decompression is no longer regarded to be accurate. Rather, nerve roots are considered to deteriorate in a continuous rather than stepwise manner.[1,2]
    - A considered approach is required; patients with acute autonomic dysfunction should undergo emergency decompression, while those with subacute or chronic dysfunction may have intervention as soon as is practicable.[3,4]
    - Emergency surgery is indicated in both patients with CESI and CESR.[5]
6. *How would you counsel the patient regarding her prognosis?*
    - The severity and duration of preoperative neurologic deficit weighs heavily on outcome.
    - Urgent decompression aims to preserve what neurologic function is still there. There is scope for postoperative improvement, but it is highly variable and uncertain. The literature is heterogeneous and sparse on data, with only small case series available. In patients with CESI, perhaps up to 90% may be able to avoid socially unacceptable bladder dysfunction if adequate decompression is achieved quickly. In patients with CESR, this may be as low as 20%.[1]
7. *Describe bladder micturition physiologic mechanisms.*
    - There are two phases: bladder filling and emptying (▶ **Fig. 123.2**).[6]
    - Bladder-filling phase:
        - The bladder accumulates increasing volumes of urine.
        - The pressure within the bladder must be lower than the urethral pressure during the filling phase.
        - Bladder filling is dependent on the intrinsic viscoelastic properties of the bladder and inhibition of the parasympathetic nerves.
        - Sympathetic nerves facilitate urine storage.
        - Sympathetic input to the lower urinary tract is constantly active during bladder filling.
        - Pudendal nerve becomes excited.
    - Bladder-emptying phase:
        - A voluntary signal is sent from the brain to begin urination and continues until the bladder is empty.
        - Bladder afferent signals ascend the spinal cord to the periaqueductal gray.
        - These afferents project to the pontine micturition center and cerebrum.
        - Once the voluntary signal to begin voiding has been issued, neurons in pontine micturition center fire maximally.
        - This causes excitation of sacral preganglionic neurons leading to the wall of the bladder to contract.
        - The pontine micturition center also causes inhibition of Onuf's nucleus leading to relaxation of the external urinary sphincter.
    - In cases of incontinence related to CES:
        - In CES, afferent and efferent nerves are both injured, and the bladder can become flaccid and distended temporarily.
        - The detrusor muscle gradually becomes spontaneously active with intermittent contractions that may cause dribbling.
        - The bladder then shrinks and its wall hypertrophies.
        - This mechanism is called *denervation hypersensitization*.

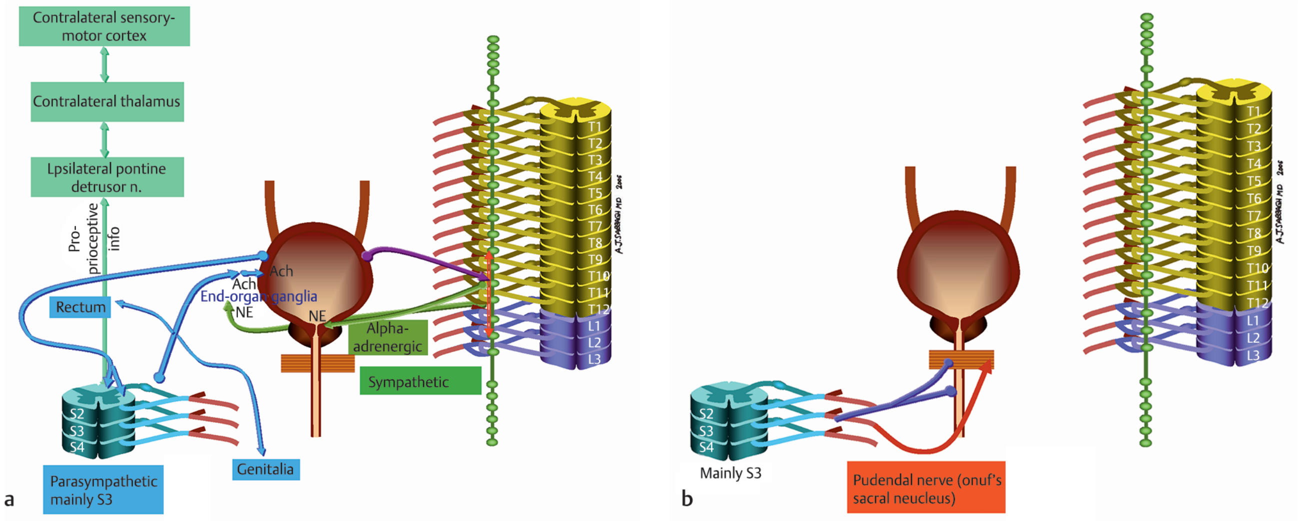
Contralateral sensory-motor cortex
Contralateral thalamus
Lpsilateral pontine detrusor n.
Pro-prioceptive info
Rectum
Ach
Ach
End-organ ganglia
NE
NE
Alpha-adrenergic
Sympathetic
T1
T2
T3
T4
T5
T6
T7
T8
T9
T10
T11
T12
L1
L2
L3
S2
S3
S4
Parasympathetic mainly S3
Genitalia
a
Mainly S3
Pudendal nerve (onuf's sacral neucleus)
b

**Fig. 123.2** Micturition physiologic mechanisms. (**a**) Bladder innervation circuits during micturition and (**b**) during relaxation phase. Ach, acetylcholine; NE, norepinephrine; N, nerve; info, information; T, thoracic; L, lumbar; S, sacral.

**Table 123.1** Common causes of cauda equina syndrome

| Type of Lesion | Etiology |
|---|---|
| Nonneoplastic compressive | Degenerative: herniated lumbosacral disc, spinal canal stenosis, facet joint cyst |
| | Epidural hematoma: iatrogenic, spontaneous |
| | Epidural abscess |
| | Traumatic fracture or traumatic disc herniation |
| Neoplastic compressive | Primary: schwannoma, ependymoma, neurofibroma |
| | Secondary: lung, breast, prostate, renal, colorectal |
| | Lymphoma |
| Noncompressive | Ischemic |
| | Inflammatory: arachnoiditis, ankylosing spondylitis |
| | Infective: HSV-2, HIV/AIDS-related infections |

*Abbreviations:* AIDS, acquired immunodeficiency syndrome; HIV, human immunodeficiency virus; HSV, herpes simplex virus.

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# Case 124 Lumbar Spondylosis with Facet Hypertrophy

*Terence Verla and Ibrahim Omeis*

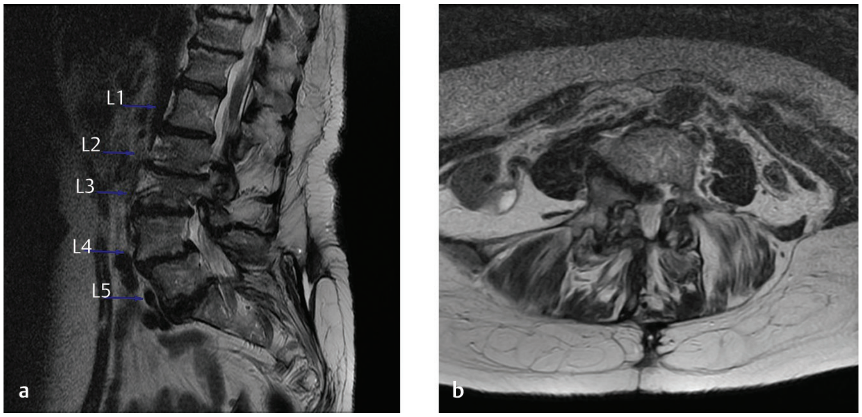

**Fig. 124.1** (**a**) MRI T2-weighted image, sagittal view of the lumbar spine. (**b**) MRI T2-weighted image, axial view of the lumbar spine.

## Clinical Presentation

- A 72-year-old female presents with chronic low back pain that radiates to her lower extremities. The pain is worse with walking or standing for extended periods of time and relieved by lying down or leaning forward. There is associated worsening numbness, tingling, and weakness in her bilateral lower extremities. She denies recent trauma, falls, bowel or bladder incontinence, or saddle anesthesia. She ambulates with the assistance of a cane.
- The patient has tried conservative therapy for at least 3 months including muscle relaxants, oral analgesics, physical therapy, and aqua therapy with no symptomatic relief.
- On examination, she has weakness in her lower extremities with Medical Research Council (MRC) grade 4+/5 in hip flexion, 4+/5 in ankle dorsiflexion, and 3/5 in great toe dorsiflexion. Sensation to light touch and pinprick was decreased in her lower extremities with no dermatomal organization. Deep tendon reflexes were 1+ for patella bilaterally and 0 in Achilles bilaterally.
- Imaging studies were ordered (▶**Fig. 124.1**).

## Questions

1. Prior to obtaining imaging studies, what is the differential diagnosis?
2. Interpret the MRI?
3. What are the different features of lumbar degenerative disc disease?
4. What is the pathophysiology and presenting symptoms of facet arthropathy?
5. Hypertrophy of which articular process (inferior or superior) results in lateral recess stenosis?
6. What are some risk factors for lumbar spondylosis?
7. What are the different treatment options?
8. What option will this patient benefit the most from?
9. Briefly describe the different surgical interventions that can be offered to this patient.

   The patient undergoes a standard multilevel laminectomy and foraminotomies for decompression of the thecal sac along with decompression of stenosed exiting nerve roots. She improves for a few months but subsequently develops progressive axial back pain that is again refractory to 3 months of conservative care. Imaging studies including MRI do not reveal any signs of instability.
10. What are your treatment options at this stage?

## Answers

1. *Prior to obtaining imaging studies, what is the differential diagnosis?*
   - Degenerative disc disease/herniation
   - Myofascial pain
   - Degenerative lumbar scoliosis with sagittal imbalance
   - Traumatic fracture
   - Spinal epidural abscess
   - Spinal cord or nerve root tumor
   - Epidural lipomatosis
   - Postviral inflammations
2. *Interpret the MRI?*
   - The MRI demonstrates severe degenerative disease in the lumbar spine in both the sagittal (▶**Fig. 124.1**a) and axial (▶**Fig. 124.1**b) T2-weighted sequences with multilevel disc height loss and facet hypertrophy causing canal stenosis.
   - There is also lateral recess stenosis compressing the traversing nerve roots and foraminal stenosis causing compression of the exiting nerve roots.
   - Bilateral facet arthropathies are appreciated at multiple levels.
3. *What are the different features of lumbar degenerative disc disease?*
   - Degeneration of the intervertebral disc results from cumulative trauma, causing disruption in the normal architecture of the disc.[1,2]
   - This is common with the aging population as loss of hydration causes a decrease in the proteoglycan content of intervertebral discs.[2]
   - This results in avulsion of the annulus fibers and subsequent herniation of the nucleus pulposus. Disruption of the annular lamellar structure decreases the ability of the disc to bear loads axially, thereby resulting in increased pressure and herniation of disc materials into the spinal canal or neural foramina.
4. *What is the pathophysiology and presenting symptoms of facet arthropathy?*
   - Facet arthropathy is a degenerative process exacerbated by disc degeneration at the same level and inflammation of the joint capsule.
   - There is subsequent wear-and-tear of the cartilaginous joint capsule.
   - The body's compensatory response is to thicken the bone around the joint, resulting in the formation of osteophytes.
   - This results in stiffness, reduced mobility of the joints, and joint pain.
   - The combination of inflammation and bone growth results in facet hypertrophy and subsequent foraminal and lateral recess stenosis.
   - Patients present with radicular pain, numbness, and tingling in the distribution of the exiting and traversing nerve roots.
5. *Hypertrophy of which articular process (inferior or superior) results in lateral recess stenosis?*
   - Superior articular process hypertrophy causes stenosis in the lateral recess zone. This, in turn, produces lateral recess stenosis impinging on the traversing nerve root.
6. *What are some risk factors for lumbar spondylosis?*
   - Nonmodifiable[3,4]:
     - Advanced age
     - Gender (males more than females)[5]
     - Hereditary/genetic factors (e.g., ankylosing spondylitis)
     - Hip osteoarthritis[6]
   - Modifiable[7,8,9]:
     - Cigarette smoking
     - Obesity
     - Sedentary lifestyle
     - Poor posture
     - Heavy weight lifting
     - Incident back trauma[5]
7. *What are the different treatment options?*
   - Nonsurgical/conservative management options:
     - Muscle relaxants, short-course analgesics, nonsteroidal anti-inflammatory drugs (NSAIDs), and oral steroids
     - Physical therapy with emphasis on strengthening core abdominal and paraspinal muscles, muscle stretching, and relaxation
     - Aerobic conditioning exercises such as walking, swimming, biking
     - Heat and ice therapy: application of heat to muscles and joints increases flexibility and range of motion. Application of ice may numb painful regions.
     - Epidural steroid injections or selective nerve root steroid injection/nerve root blocks
     - Weight lifting limitations, intermittent use of a back brace
     - Smoking cessation and weight loss
     - Other lifestyle modifications, postural adjustments
     - Alternative treatment options may be used within a limited scope depending on the severity of the condition, these include: chiropractic adjustments, acupuncture, acupressure, aquatic therapy, Alexander technique, Rolfing techniques, relaxation and stretching exercises, yoga, tai chi.
   - Surgical indications
     - Failure of conservative management options over at least 6- to 12-week period
     - Severe/disabling neurogenic claudication
     - Progressive worsening in motor strength
     - Global sagittal imbalance
     - Signs of cauda equina syndrome including bowel/bladder incontinence or saddle anesthesia

## Answers (*continued*)

8. *What option will this patient benefit the most from?*
   - The patient has already tried 3 months of physical therapy and other conservative care with minimal improvement in symptoms.
   - She is having worsening of her lower extremity weakness.
   - She now requires a cane to ambulate.
   - Therefore, the patient could likely benefit from surgical intervention, should there be no contraindications for her to undergo a surgical procedure.
9. *Briefly describe the different surgical interventions that can be offered to this patient.*
   - When treating patients with pure neurogenic claudication and minimal back pain, surgical decompression alone can be offered. Surgery can be focal to address the specific pathologic level causing the symptoms, or, if the disease is multilevel, a longer decompression may be appropriate.
   - When patients have severe low back pain and sagittal imbalance with degenerative lumbar scoliosis, in addition to neurogenic claudication, a more extensive surgical procedure is typically required that usually entail a multilevel decompression, with restoration of lumbar lordosis and instrumented fusion. The patient undergoes a standard multilevel laminectomy and foraminotomies for decompression of the thecal sac along with decompression of stenosed exiting nerve roots. She improves for a few months but subsequently develops progressive axial back pain that is again refractory to 3 months of conservative care. Imaging studies including MRI do not reveal any signs of instability.
10. *What are your treatment options at this stage?*
   - The patient has developed a postlaminectomy syndrome that is a well-recognized disease entity. The treatments of postlaminectomy syndrome include physical therapy, low-force specific chiropractic care, nerve blocks, transcutaneous electrical nerve stimulation (TENS), behavioral medicine, NSAIDs, membrane stabilizers, and antidepressants.
   - In cases where conservative treatments fail, spinal cord stimulation, and intrathecal morphine pump remain the mainstay of treatment and are considered “last resort” options.
   - Use of epidural steroid injections may be minimally helpful in most cases.

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# Case 125 Degenerative Spondylolisthesis

*Ralph J. Mobbs, Monish Maharaj, and Kevin Phan*

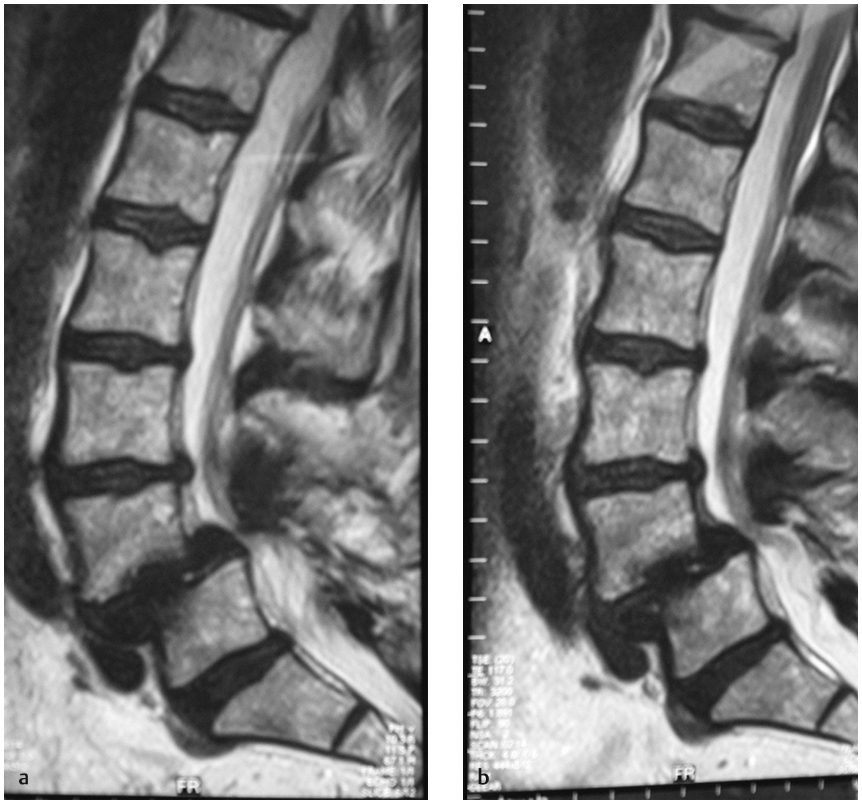

**Fig. 125.1** View through median sagittal plane MRI revealing grade 3 degenerative spondylolisthesis at the L4–L5 level with secondary cord compression.

## Clinical Presentation

- A 63-year-old woman presents in a wheelchair bound state following severe lower back pain that has lasted over 5 years. Conservative bed rest, medication use, and bracing have been attempted unsuccessfully throughout this period. She is unable to walk as this exaggerates the pain.
- Past history reveals a thoracic epidural abscess 6 years ago, with a laminectomy procedure 4 years ago.
- There is no specific spinal tenderness noted, but bilateral limitations in power and movement across all lower limb myotomes are noted.
- An MRI of the patient is obtained (see ▸**Fig. 125.1**).

## Questions

1. Describe the MRI (▸**Fig. 125.1**).
2. What imaging modalities could be considered in the work-up for this patient?
3. Briefly describe the anatomical structures that reinforce the lumbar interspinal joint and the underlying pathological issue in the setting of degenerative spondylolisthesis.
4. How is spondylolisthesis classified? What type does this patient most likely belong to?
5. What signs and symptoms may the patient present with that reinforce a diagnosis of spondylolisthesis?
6. What treatment options are recommended for patients with conservative treatment-resistant degenerative spondylolisthesis?
7. Does the presence of multilevel disease alter the treatment recommendations you would offer to the patient?

## Answers

1. *Describe the MRI (▶**Fig. 125.1**).*
   - Views through the median sagittal plane T2-weighted MRI demonstrate grade 3 (Taillard method) spondylolisthesis involving the L4–L5 vertebrae with associated neural elements compression. The L4–L5 level is the most frequently affected level in the setting of degenerative spondylolisthesis.
   - An axial view would show diffuse disc bulge, ligamentum flavum hypertrophy, and bilateral facet joint arthritis with consequent spinal canal stenosis.
   - Considering the patient's age and the long-standing deterioration, the most probable diagnosis is spondylolisthesis, likely degenerative in nature. The findings are consistent with a grade 3, type III spondylolisthesis.
2. *What imaging modalities could be considered in the work-up for this patient?*
   - Radiographs: Anteroposterior and lateral view plain radiographs are recommended in patients with low back pain if over 50 years of age, or those demonstrating other systemic signs, and sciatica with radiculopathy. In the setting of severe acute deterioration or progressive motor weakness, more sophisticated imaging should be sought.
     - In the setting of degenerative spondylolisthesis, radiographs may demonstrate arthritic changes surrounding the facet joints.
     - Although statistically significant differences in slippage magnitude is noted across plain standing or recumbent radiographs or flexion-extension radiographs, the former is shown to demonstrate a greater magnitude of sagittal translation, making diagnosis more likely.[1]
   - MRI: Although not as sensitive for bony pathology, in the setting of spondylolisthesis MRI is useful for the evaluation of spinal stenosis, nerve root compression, as well as the condition, contents, and integrity of the disc tissue. The presence of significant facet joint effusion (> 1.5 mm) is suggestive of degenerative spondylolisthesis.[2] It should be considered in the setting of patients with treatment-refractory radicular pain or those with abnormal radiographic findings.
   - CT: It remains the gold standard for the diagnostic detection of bony abnormalities of the spine including spondylolisthesis and may be useful when radiographs are abnormal or inconclusive. CT may illustrate the signs of facet joint arthritis including increased enhancement at the site.
     - However, limitations include costs, radiation exposure, and a limited ability for the radiologist to distinguish between other tissues such as scar, ligaments, and the spinal discs.
   - Additional studies, such as electromyelography (EMG), discography, and bone scanning, have controversial roles and are often not necessary in the setting of degenerative spondylolisthesis when MRI is readily accessible. EMG response may be of use in evaluating good surgical candidates, but is not essential in establishing a diagnosis.
3. *Briefly describe the anatomical structures that reinforce the lumbar interspinal joint and the underlying pathological issue in the setting of degenerative spondylolisthesis.*
   - Like the smaller thoracic vertebrae, the body of the lumbar segments are concave from above down and may be wedge-like in shape with pedicles originating from the upper half and a near horizontal spinous process.
   - Each unit is reinforced by the anterior and posterior longitudinal ligaments running from the base of the skull to the sacrum, linking the bony structures with the annulus fibrosis. The ligamentum flavum is contained over the posterior aspect of the spinal canal, while the supraspinous and interspinous ligaments reinforce the vertebral segments posteriorly via the spinous processes.
   - Finally, the upper four transverse processes attach directly to the psoas fascia and the anterior layer of the lumbar fascia, facilitating the stabilization of the spine.[3]
   - The symptoms present are primarily due to a combination of reduction in sagittal diameter and stenosis of the neural foramen. In the setting of degenerative spondylolisthesis, this may present as classical osteoarthritic features of the facet joints of the slipped vertebra including osteophyte formation, joint subluxation, and loss of joint space. This instability may also cause tension to the joint capsule and ligaments that may lead to stabilizing muscle overuse and ligament fibrosis.[4]
   - The presence of a bulging disc may compound to the symptoms experienced.[5]
4. *How is spondylolisthesis classified? What type does this patient most likely belong to?*
   - There are two broad classifications for spondylolisthesis, one evaluating the underlying etiology (Wiltse criterion) and the other evaluating the degree of slippage (Taillard method, see ▶**Fig. 125.3**). Both have implications on treatment.
   - The Wiltse classification segregates spondylolisthesis based on etymology. It is broadly categorized in to seven groups, although subtypes exist. In order they include, dysplastic/congenital, isthmic, degenerative, posttraumatic, pathologic, and iatrogenic.[6]
   - The Taillard method of grading slippage is generally accepted as the most reproducible, expressing the degree of slippage between the anteroposterior diameter and the top of the inferior segment. Grades 1 to 4 are simply based on the percentage of slippage. A grade of 3 or 4 (> 50% slippage)

generally requires surgical intervention. Notably, interobserver error rates of up to 15% have been reported, with the number exaggerated in the presence of segment rotation.[4]

- Newer methods of classification have been proposed but are not yet routinely used in practice.[7,8]

5. *What signs and symptoms may the patient present with that reinforce a diagnosis of spondylolisthesis?*
   - The diagnosis of degenerative spondylolisthesis is primarily radiological. However, the most commonly reported symptoms include occasional or chronic lower back pain. Patients may or may not describe radiculopathy, positional variation to pain, or intermittent claudication.
   - These nonspecific symptoms may also be explained by other diagnoses including disc prolapse, reiterating the importance of diagnostic imaging.[5]
6. *What treatment options are recommended for patients with conservative treatment-resistant degenerative spondylolisthesis?*
   - For patients with conservative treatment-resistant degenerative spondylolisthesis with spinal stenosis, surgical intervention remains the mainstay of therapy. Conservative options include medication use, physical rehabilitation, bracing, spinal exercises, and manipulation as well as combination therapy.[9]
   - Decompression with fusion is suggested for patients with high-grade disease.
   - In patients with low-grade slippage (< 20%) current North American Spine Society (NASS) guidelines advise for decompression with midline structure preservation, as outcomes are generally comparable to those receiving decompression with fusion.[2]
   - There is limited data regarding the best approach for fusion, although randomized comparative studies have demonstrated superior outcomes in objective pain measures associated with transforaminal lumbar interbody fusion (TLIF) and LLIF in comparison to a posterior fusion (▶**Fig. 125.2**).[10–12] There is no general consensus regarding the best approach for fusion as patient factors and the level of disease may affect the choice of method.[13] The presence of an incidental durotomy during the procedure does not appear to affect outcomes at 12-month follow-up.[14]
   - The use of further instrumentation, circumferential fusion, and minimally invasive approaches remain topics of ongoing research.
7. *Does the presence of multilevel disease alter the treatment recommendations you would offer to the patient?*
   - Multilevel degenerative lumbar spine disease may involve other factors of disease such as deformity or additional disc herniation that may also require correction through operative means.
   - There are low-powered studies demonstrating superior clinical outcomes with minimally invasive TLIF compared to a conventional posterior lumbar interbody fusion (PLIF) in the setting of three-level disease; however, the choice of surgical approach and procedure is ultimately dictated by the individual patient characteristics.

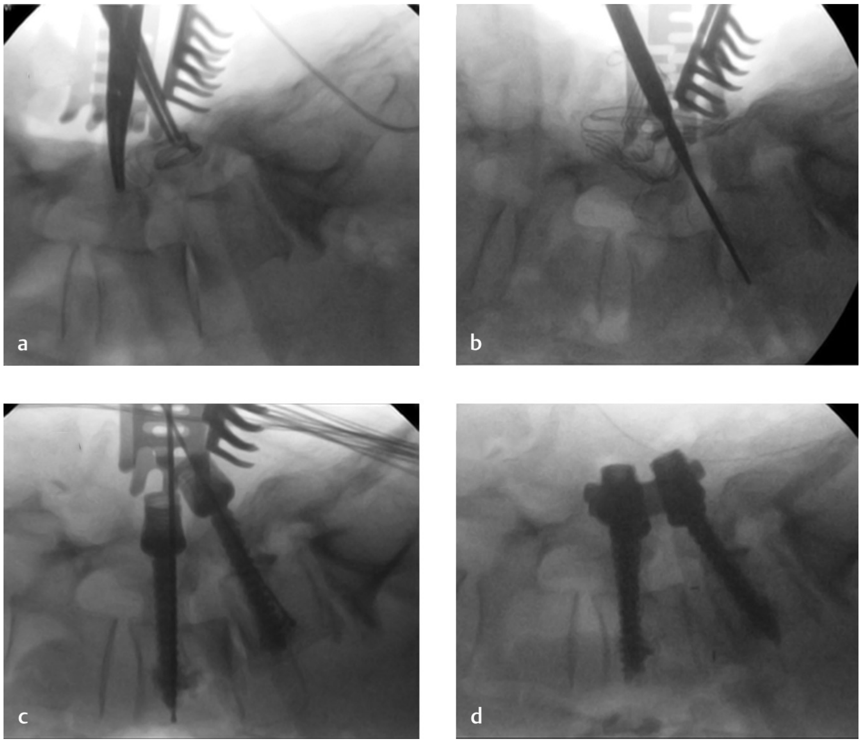

**Fig. 125.2** (**a**) Initial exposure and identification of transverse process of L4 and L5. (**b**) Laminectomy and insertion of interbody distraction tool to prepare endplates. (**c**) Insertion of pedicle screws. Note: Cement used to assist with firm fixation for reduction maneuver. (**d**) Cage insertion and reduction maneuver.

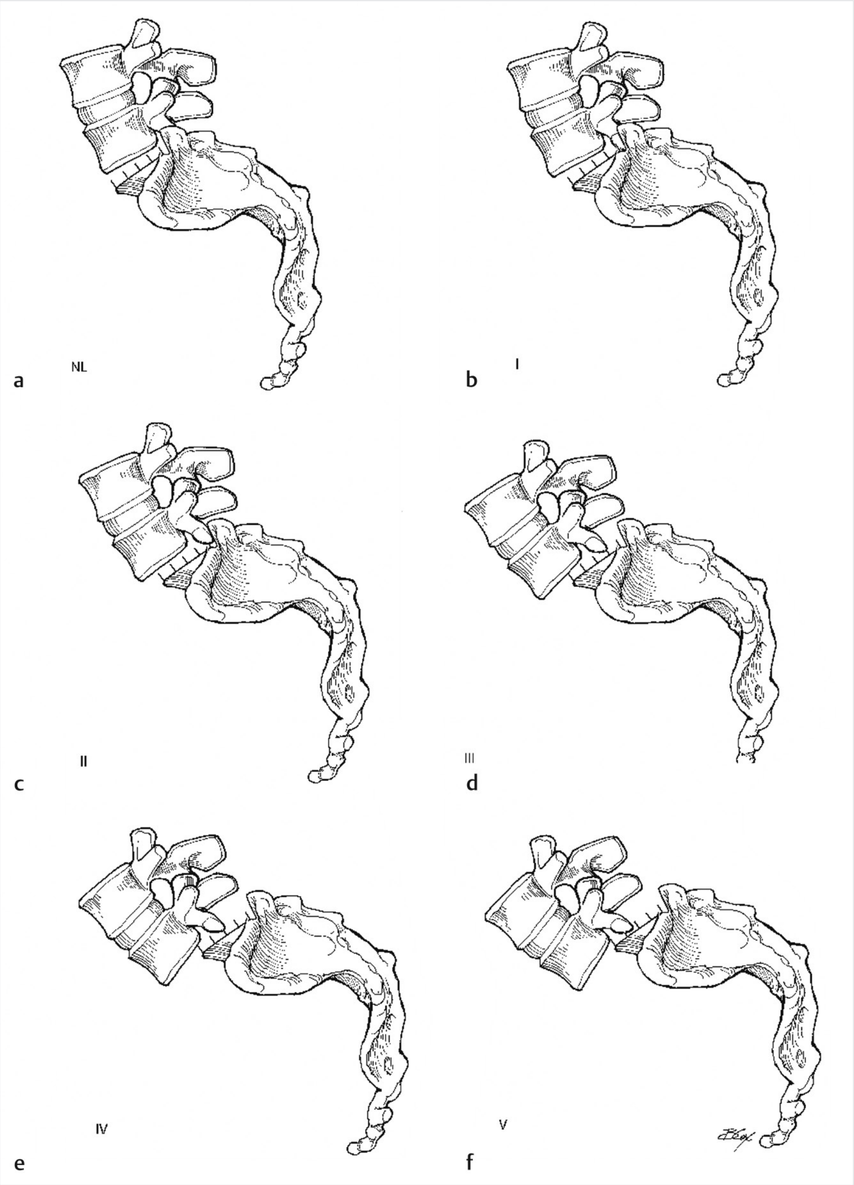

**Fig. 125.3** Classification of spondylolisthesis: Normal, Grade I, 0 to 25%; Grade II, 26 to 50%; Grade III, 51 to 75%; Grade IV, 76 to 100%; Grade V, >100% (spondyloptosis). (Source: 25.3.5 Spondylolisthesis. In: Baaj A, Mummaneni P, Uribe J et al., ed. Handbook of Spine Surgery. 2nd Edition. Thieme; 2016.)

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# Case 126 Sacroiliac Joint Dysfunction SIJ Fusion

*Mohammad Almubaslat, Anup Aggarwal, Cristian Gragnaniello, and Remi Nader*

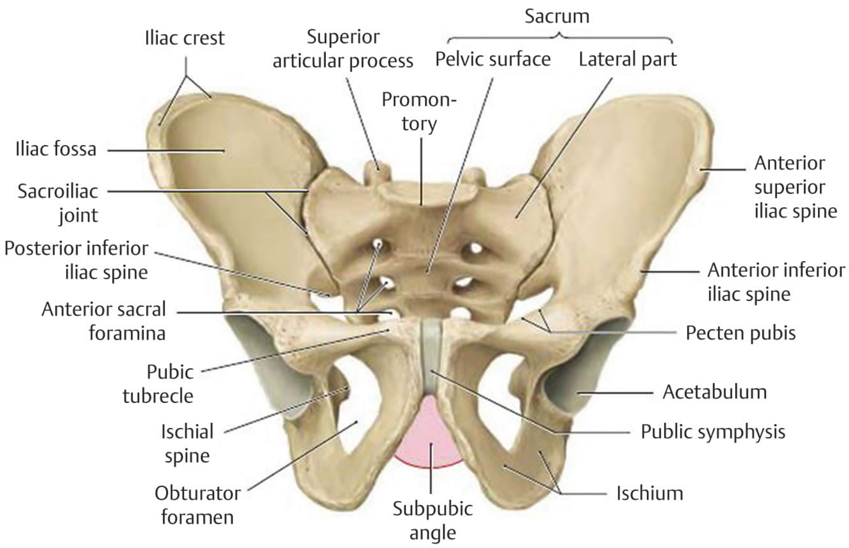
Sacrum
Iliac crest
Superior articular process
Pelvic surface
Lateral part
Promontory
Iliac fossa
Sacroiliac joint
Anterior superior iliac spine
Posterior inferior iliac spine
Anterior inferior iliac spine
Anterior sacral foramina
Pecten pubis
Pubic tubrecle
Acetabulum
Ischial spine
Public symphysis
Obturator foramen
Ischium
Subpubic angle

**Fig. 126.1** Sacroiliac joint anatomy. Anterior view. (Reproduced from Schuenke M et al. Thieme Atlas of Anatomy General Anatomy and Musculoskeletal System. New York: Thieme; 2010. Illustration by Karl Wesker/Markus Voll.)

## Clinical Presentation

- A 58-year-old woman presents with progressive right-sided back pain and right leg pain of 2-year duration. She has a history of lumbar fusion 10 years prior at L4–S1. She describes the pain as sharp and stabbing in the right buttock and hip, radiating to the right anterior and lateral thigh to the knee. She has no weakness or sensory symptoms. She has difficulty finding a good sitting position, and laying on her right buttock exacerbates the pain.
- She had three lumbar epidural steroid injections at and above the fusion levels by a pain management specialist, but only responded mildly to the first injection. She also underwent physical therapy for 2 months on two different occasions without improvement.

## Questions

1. What are the pertinent areas and systems that should be examined in this patient?
2. Describe the basic anatomy of the sacroiliac joint (SIJ).
3. What are the different degrees and directions of motion of the SIJ?
4. What is SIJ dysfunction?
5. What are the factors that favor the diagnosis of SIJ dysfunction versus lumbar radiculopathy in the current patient?
6. What are the signs and provocative tests used to identify SIJ dysfunction on physical examination?
7. What are the clinical diagnostic criteria for establishing SIJ dysfunction and initiating treatment?
8. What are some of the nonsurgical treatment options for SIJ dysfunction?
9. What are the surgical treatment options for SIJ dysfunction?
10. What are the clinical diagnostic guidelines for proceeding with surgical treatment of SIJ dysfunction with arthrodesis?
11. Describe briefly the minimally invasive SIJ fusion procedure.
12. What are some of the postoperative complications associated with minimally invasive SIJ fusion?

## Answers

1. *What are the pertinent areas and systems that should be examined in this patient?*
   - Musculoskeletal examination of the spine
   - Neurological exam
   - Musculoskeletal examination of the SIJ
2. *Describe the basic anatomy of the sacroiliac joint (SIJ).*
   - The SIJs are paired joints that are formed between the articular surfaces of the sacrum and the iliac bones. The sacral surface is covered with hyaline cartilage, whereas the ilium has fibrocartilage. The joint surface starts flat early in life, but develops irregularities and crevices as we age. These irregularities, coupled with a strong intrinsic and extrinsic complex of ligaments lead to high stability in the joint, allowing only a small degree of motion between 2 to 18 total degrees of motion in various directions. The degree of motion is higher in women than men, which accommodates pelvic changes during pregnancy and delivery.
   - The main innervation of the SIJ area is from the dorsal lateral sacral nerve roots. Innervation ranges from L5 to S4, but in the majority of cases the SIJ is innervated from S1 to S3 nerve roots (see ▸**Fig. 126.1**).
3. *What are the different degrees and directions of motion of the SIJ?*
   - Anterior tilt of both iliac bones on the sacrum (moving as a unit)
   - Posterior tilt of both iliac bones on the sacrum
   - Anterior tilt of one iliac bone with simultaneous posterior tilt of the other side on the sacrum (antagonistic tilt), which occurs during gait
   - Sacral flexion (or nutation) which describes rotation-translation displacement, or forward tilt of the sacrum relative to the ilia deepening on the lumbar lordosis
   - Sacral extension (or counter-nutation) which describes the opposite motion, decreasing the lumbar lordosis
4. *What is SIJ dysfunction?*
   - SIJ dysfunction, is also referred to as SIJ disorder, SIJ disease, SIJ syndrome, or sacroiliitis (although a true inflammatory process only occurs in certain pathological conditions, the term sacroiliitis has been used as an umbrella term). SIJ dysfunction generally refers to pain in the SIJ region that is caused by abnormal motion in the joint, due to either *hypermobility* or *hypomobility*. It is estimated to be prevalent in 15 to 30% of all chronic low back pain patients, and up to 61% of postlumbar fusion patients due to increased biomechanical stress on both SIJ after lumbar fusion.
   - *Hypermobility* is thought to be the type that causes the majority of cases of SIJ dysfunction. In these cases, the dysfunction is usually extra-articular because abnormal joint movement and alignment is a consequence of weakened, injured, or sprained ligaments around the joint. These can be caused by degeneration, direct iatrogenic causes (after extensive bone harvesting from the iliac bone), indirect iatrogenic causes (such as altered biomechanics after spinal surgery), or less common causes, such as infection, neoplasm, or trauma leading to fracture or dislocation.
   - *Hypomobility*, on the other hand, occurs due to intrinsic abnormalities in the joint associated with high irregularities in the joint articulating surfaces. These can result from a variety of conditions, such as advanced degeneration, old trauma, abnormal scarring, remote infection, neoplasm, or inflammatory conditions, such as ankylosing spondylitis or rheumatoid arthritis. In ankylosing spondylitis, the inflammatory process is aggressive and can lead to complete erosion of the joint surfaces leading to an autofusion phenomenon of both joints, which is the hallmark of this condition.
5. *What are the factors that favor the diagnosis of SIJ dysfunction versus lumbar radiculopathy in the current patient?*
   - MRI scan showing no significant central or lateral stenosis, lumbar instability, or significant disc disease, especially at levels adjacent to any prior fusion.
   - Sensory or motor deficit that does not follow a typical radicular pattern.
   - Previous history of lumbar fusion
   - Previous history of bone graft harvest from the iliac crest, usually from the symptomatic side.
   - Clear radiographic evidence of SIJ degeneration, such as sclerosis, osteophytes, subchondral cysts, vacuum phenomenon, or other pathological conditions related to infection, trauma, or neoplasm.
   - Positive clinical signs on examination that are pathognomonic of SIJ disease (see below)
   - See ▸**Fig. 126.2**, ▸**Fig. 126.3**, and ▸**Fig. 126.4** for illustrative case
6. *What are the signs and provocative tests used to identify SIJ dysfunction on physical examination?*
   - Fortin's sign: When the patient is asked to point the maximal area of pain with a finger, the patient points 1 cm inferior and medial to the posterior superior iliac spine (PSIS) on the symptomatic side on two separate occasions.
   - Provocative tests:
     - Pelvic gapping (distraction): In supine position, application of posterior pressure on both anterior superior iliac spines (ASIS) points simultaneously causes pain in the SIJ area posteriorly.
     - Compression test: In lateral recumbent position, application of pressure directly down on the

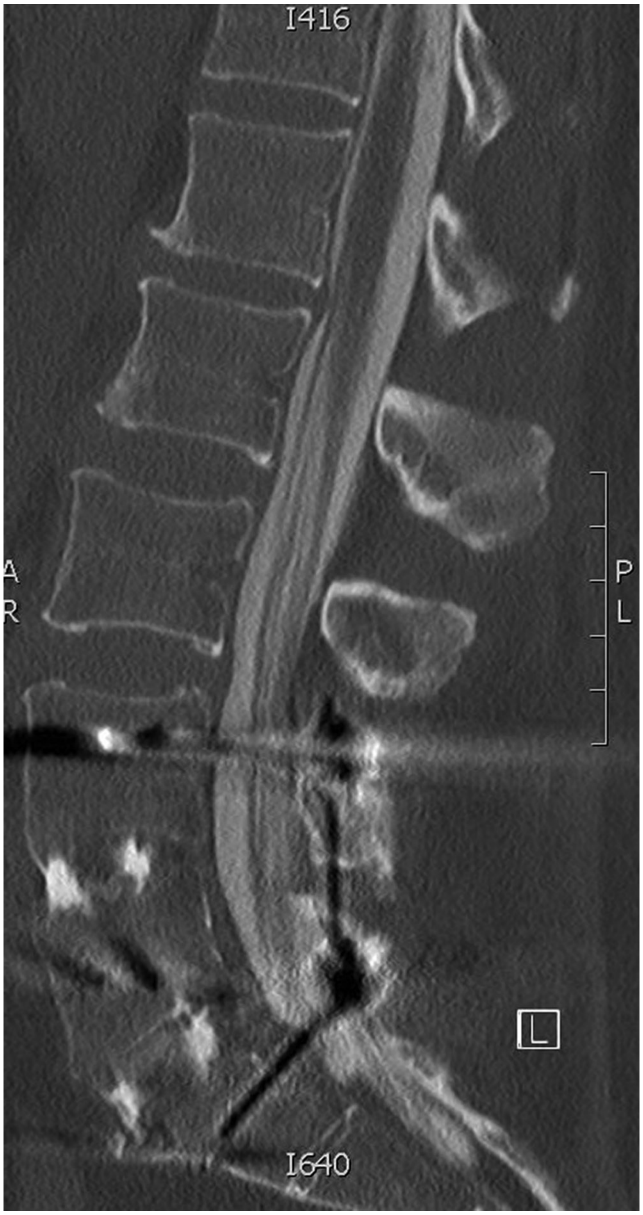

**Fig. 126.2** Sagittal CT myelogram of the patient showing previous lumbar fusion. No significant stenosis was found in the lumbar spine.

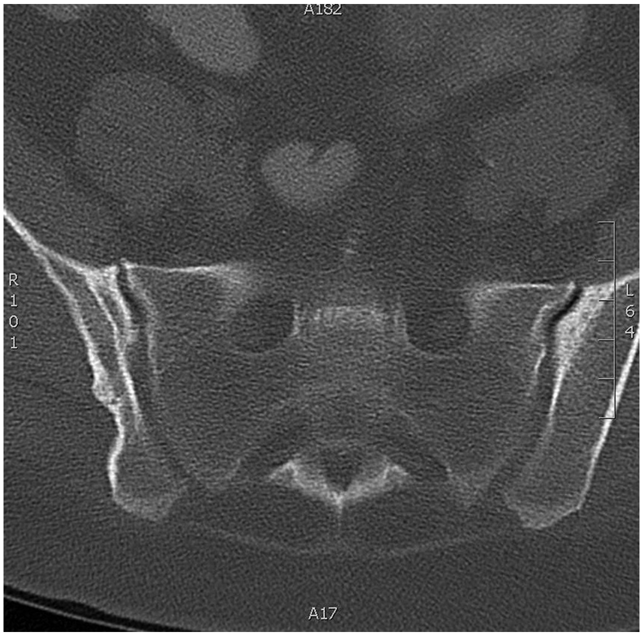

**Fig. 126.3** Axial CT of the sacroiliac joints revealing abnormal thinning and ridging of the right iliac bone secondary to previous bone graft harvesting.

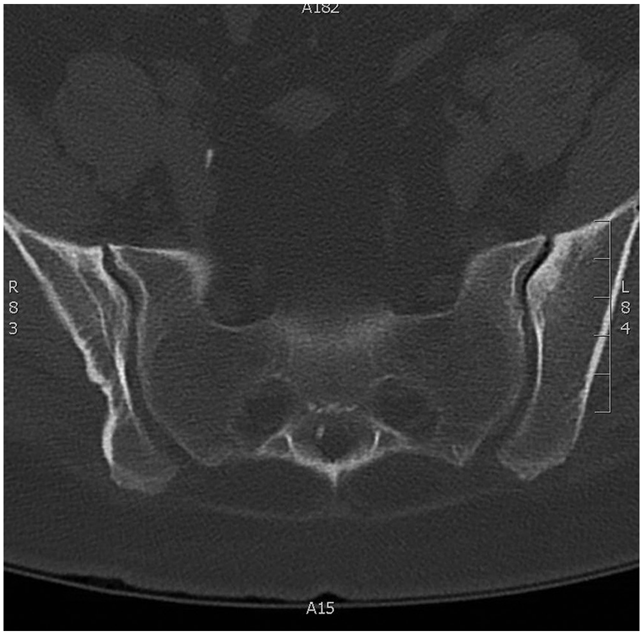

**Fig. 126.4** Axial CT of the sacroiliac joints, different level, also showing the right iliac abnormality close to the SI joint.

## Answers (*continued*)

superior hip's lateral aspect causes pain in the SIJ area posteriorly.
  - Flexion, abduction, external rotation (FABER) test: In supine position, application of flexion, abduction, and external rotation of the leg on the affected SIJ side, while stabilizing the contralateral side, causes pain in the affected SIJ area.
  - Thigh thrust: In supine position, with the contralateral hip flexed and stabilized, application of pressure down on the ASIS of the ipsilateral (affected) side causes pain in the affected SIJ area.
  - Gaenslen's test: In lateral recumbent position, the contralateral hip is flexed maximally, and the ipsilateral or affected hip is extended while maintaining stability of the pelvis; this causes pain in the affected SIJ area.

7. *What are the clinical diagnostic criteria for establishing SIJ dysfunction and initiating treatment?*
   - History of SIJ-localized pain: SIJ pain score ("average SIJ pain in the last week") of at least 5 on a 0 to 10 visual analog scale (VAS), where 0 represents no pain and 10 represents the worst pain the patient ever had in their life.
   - Positive provocative testing on at least three of the five established tests (see ▶**Fig. 126.5**):
     - SIJ distraction
     - SIJ compression
     - FABER test
     - Thigh thrust
     - Gaenslen's test
8. *What are some of the nonsurgical treatment options for SIJ dysfunction?*
   When the initial clinical diagnostic criteria are established, one of the following treatment modalities can be used:
   - Oral pain medications
   - Physical therapy (formal stretching exercises, postural modification, and lifestyle adjustments)
   - Manipulative therapy (e.g., chiropractic treatments and osteopathic adjustments)
   - Use of sacral orthotic devices: thought to reduce motion at the joint
   - SIJ injections (e.g., steroids and/or local analgesics such as lidocaine; autologous stem cell and platelet rich plasma injections have also been tried with some success although further research is needed to substantiate these techniques).
   - Radiofrequency ablation (RFA): Studies of RFA of sacral nerve root lateral branches showed short-term reduction in SIJ pain (12 months), and more moderate long-term gain.
9. *What are the surgical treatment options for SIJ dysfunction?*
   Since most cases of SIJ dysfunctions are cases associated with *hypermobility*, it was proposed that SIJ arthrodesis would lead to improvement or elimination of SI pain.
   - Open SIJ arthrodesis with screws and plates has been used for years, especially in traumatic cases.
   - Minimally invasive SIJ fusion using fusion screws or triangular titanium implants: more contemporary procedure that was described in the past 15 years.
10. *What are the clinical diagnostic guidelines for proceeding with surgical treatment of SIJ dysfunction with arthrodesis?*
    - Establishment of the clinical diagnostic criteria described above.
    - Failure of other nonsurgical treatment methods: No established guidelines, but it has been proposed to try these modalities for 6 months before considering surgery.
    - Performing a diagnostic SIJ injection (typically of lidocaine):
      - At least a 50% decrease (but preferably 75%) in SIJ pain 30 to 60 minutes after image-guided local anesthetic injection (utilizing contrast) into the symptomatic SIJ performed within 3 months of consideration for surgery.
    - Obtaining diagnostic imaging: X-ray, CT, or MRI:
      - These are used to identify presence of significant degenerative signs, such as evidence of sclerosis, osteophytes, subchondral cysts, vacuum phenomenon, or to rule out other disease processes, such as significant infection, SIJ disruption, presence of neoplasm or fracture dislocation. Note that presence of radiographic evidence of degeneration is not required for the diagnosis of SIJ dysfunction, since most cases with hypermobility are due to extra-articular dysfunction.
      - SIJ disruption manifests as pain in the context of asymmetric widening of SI joints on CT or X-rays, or the presence of significant contrast leakage during a diagnostic SIJ block.
      - One should discuss the image findings with the radiologist and make sure they are documented. Many radiologists do not make it a habit to comment on the SIJ when interpreting a pelvic CT, and will often only comment on the internal pelvic organs instead.
11. *Describe briefly the minimally invasive SIJ fusion procedure.*
    - In these procedures, the patient is placed in a prone position on a spinal radiolucent table. One or two fluoroscopic C-arms are positioned in anteroposterior (AP) and lateral positions. The AP position is used to obtain pelvic inlet and outlet views.
      - The pelvic inlet view is directed parallel to the sacrum, and is used to assure that the implant is not violating the pelvis at the most anterior border.
      - The pelvic outlet view is aimed perpendicular to the sacrum and is used to view the neural forami-

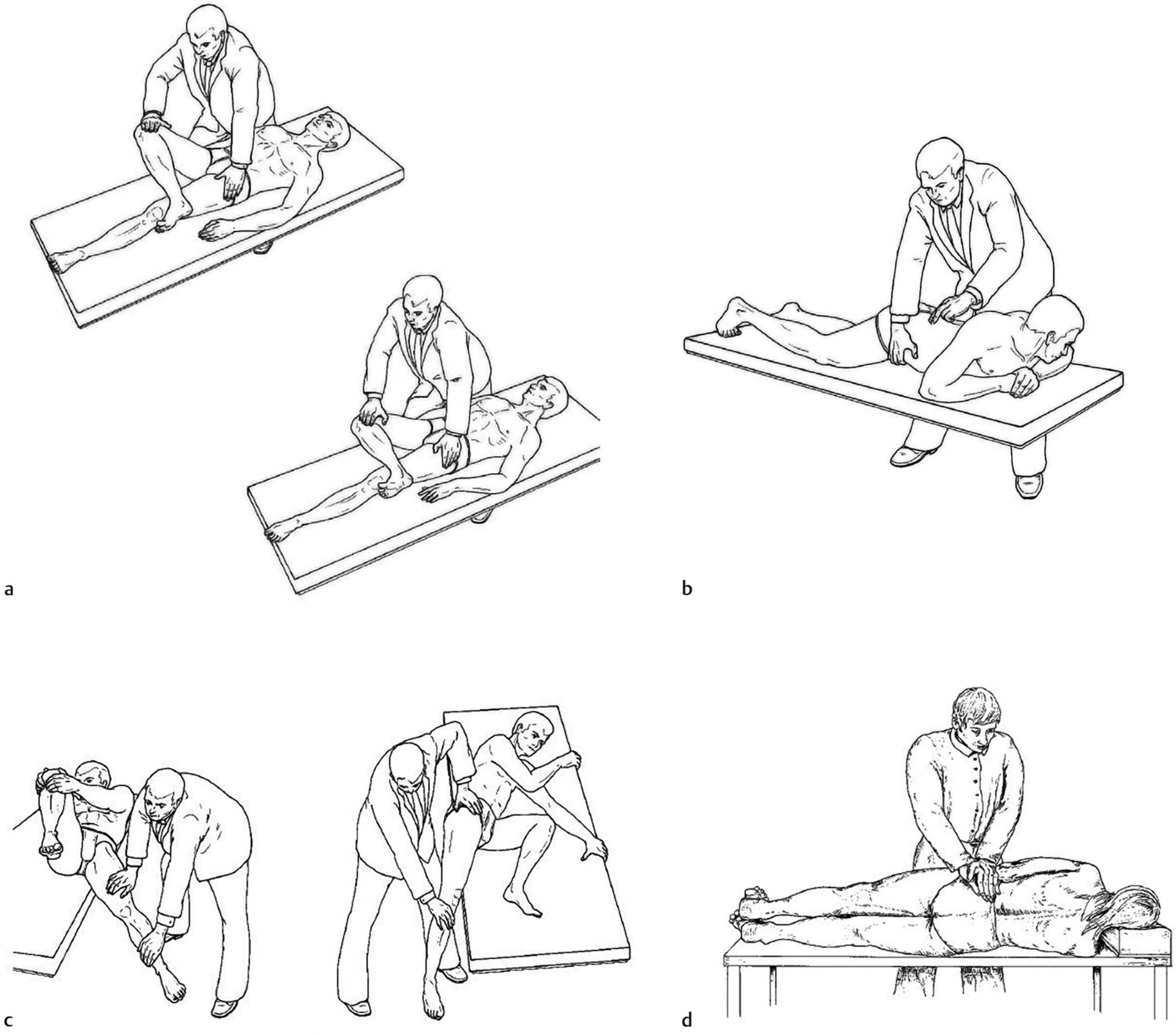

**Fig. 126.5** Illustration of some provocative tests used in the diagnosis sacroiliac joint dysfunction. (**a**) FABER or Patrick test (flexion, abduction, and external rotation). (**b**) Sacroiliac mobilization test. (**c**) Gaenslen's test. (**d**) Sacroiliac compression. (Reproduced from Buckup K. Clinical Tests for the Musculoskeletal System. 2nd ed. New York: Thieme; 2008).

## ■ Answers (*continued*)

  na and assure the implants are not violating the neural foramina.
  - The lateral view is used to identify the superior limit of the SIJ as well as its rosrtocaudal extent when selecting an entry point for the implant.
- The incision is made on the side of the gluteal area under lateral fluoroscopic visualization, and minimally invasive technique is used to place the implants. The implants are placed over a K-wire or Steinmann pin that is directed along the correct trajectory in all three planes of fluoroscopy (inlet, outlet, and lateral views).
- Typically, one to three screws or two to three triangular implants are placed depending on the particular system used. Fusion screws usually allow for placement of bone graft inside, whereas the triangular implants are coated with porous titanium plasma spray to allow growth of the bone into the side of the implant.
- See ▶**Fig. 126.6** for illustrative X-ray.

12. *What are some of the postoperative complications associated with minimally invasive SIJ fusion?*
    - Postoperative hematoma: Can be identified clinically or via CT scan. Treatment: observation, if pain continues to increase, consider surgical evacuation, exploration for vascular injury, placement of drain.
    - Lumbosacral radiculopathy: CT scan is indicated to assess violation of any of the sacral neural foramina,

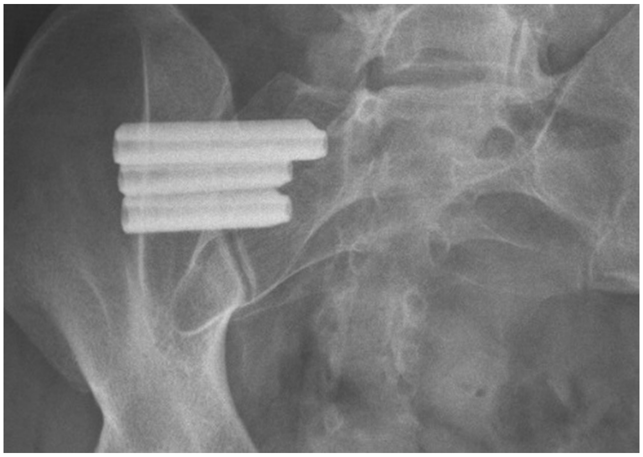

**Fig. 126.6** Postoperative X-ray of the pelvis after minimally invasive sacroiliac joint fusion using triangular implants (iFuse system from SI Bone).

## Answers (*continued*)

or compression of the L5 nerve root due to unrecognized sacralization of an L5 transitional vertebrae. This may also present as new-onset incontinence. Treatment: conservative initially if the implant is in adequate position; reoperation to modify the implant position if necessary.

- Persistent pain despite technically successful surgery: It is essential to reassess surgical indications and other confounding factors in such case as the pain may be multifactorial. One should also consider the possibility of misdiagnosis.
- Abdominal organ injury or visceral injury from anterior trajectory of implant: CT scan is indicated based on the clinical assessment.
- Vascular injury: rare; may require the assistance of a vascular surgeon for management.
- Superficial or deep wound infection
- Fracture of the ilium: reported at the inferior edge of the ilium. Usually nondisplaced. Treatment is usually conservative.

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# Case 127 Intradural Spinal Tumor

*Adam Sauh Gee Wu and Stephen J. Hentschel*

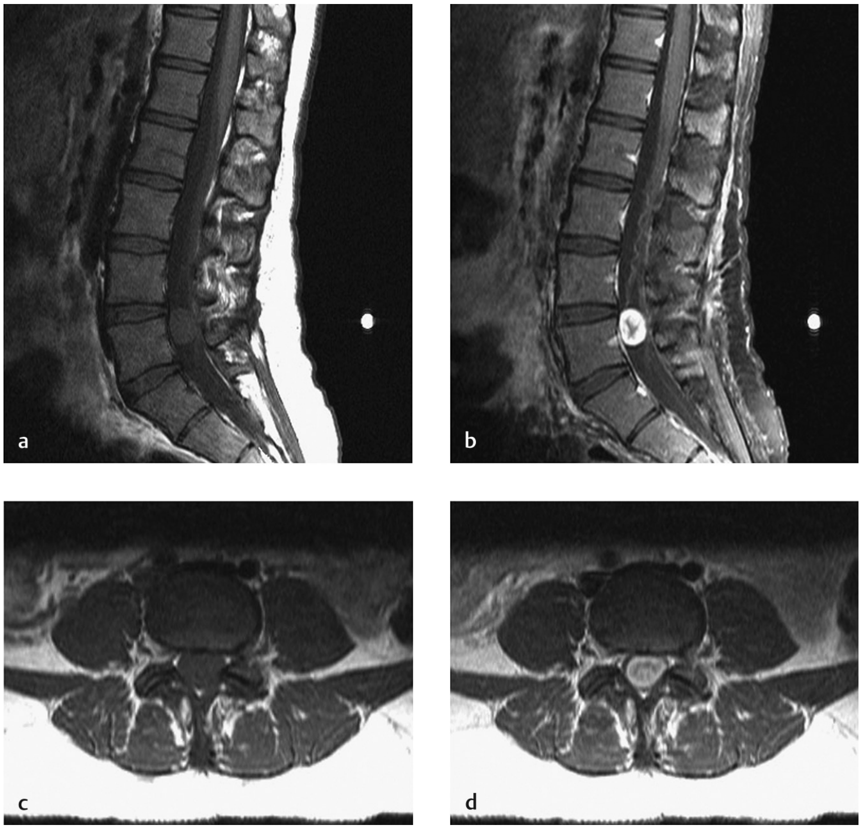

**Fig. 127.1** MR images of the lumbar spine. (**a**) Sagittal T1-weighted image, (**b**) sagittal T1-weighted image with gadolinium, (**c**) axial T1-weighted image, (**d**) axial T1-weighted image with gadolinium through the lesion at L5.

## Clinical Presentation

- A 25-year-old woman presented with 1-year history of persistent bilateral leg pain radiating from the low back to the posterior thigh and lateral ankle.
- The pain is more severe on the left side than it is on the right.
- Physical examination reveals decreased pinprick sensation over the left lateral leg and ankle. Otherwise, she is neurologically intact.
- A noncontrast CT scan of the lumbar spine was obtained and was within normal limits.

## Questions

1. Is further imaging necessary? What are you looking for?
2. An MRI scan is obtained (▸**Fig. 127.1**). Describe the findings and exact location of the abnormality.
3. What is your differential diagnosis?
4. What are the indications for and goals of surgery in this case? Is there any role for nonsurgical management?
5. Describe the surgical procedure you would perform. What intraoperative findings would you expect with respect to the relationships to neural structures and the site of origin of the mass?
6. What operative adjuncts might you consider?
7. What are the potential complications of surgery in this case, and what can you do during surgery to reduce the risk of complications?
8. Hematoxylin and eosin stains of the specimen at low and high power are shown (▸**Fig. 127.2**). Describe the histopathology. What is the diagnosis?
9. Describe the cytogenetics associated with this tumor.
10. What is the expected outcome of surgery in this case, and what is the potential for recurrence?

## Answers

1. *Is further imaging necessary? What are you looking for?*
   - Although the CT scan is normal, an MRI scan is needed to rule out intradural pathology.
2. *An MRI scan is obtained (▶***Fig. 127.1***). Describe the findings and exact location of the abnormality.*
   - There is an intradural mass at the L5 level.
   - It is isodense on T1-weighted images, with intense gadolinium enhancement but no obvious dural tail.
3. *What is your differential diagnosis?*
   - The most common intradural neoplasms of the spinal cord and cauda equina with the MRI characteristics seen in this case are meningiomas, schwannomas, neurofibromas, and myxopapillary ependymomas.
   - Metastases, hemangiopericytomas, and lipomas are less likely.[1,2,3]
4. *What are the indications for and goals of surgery in this case? Is there any role for nonsurgical management?*
   - The indications for surgery are cytoreduction to relieve mass effect, and to obtain tissue for pathologic analysis.[1,4]
   - The goal of surgery is complete resection of the tumor with preservation of neurologic function,[1] but subtotal resection is preferred over aggressive tumor removal, which risks neurologic injury, as most tumors are benign.
   - Nonsurgical management may be considered if the patient has high surgical risk or limited life expectancy.
5. *Describe the surgical procedure you would perform. What intraoperative findings would you expect with respect to the relationships to neural structures and the site of origin of the mass?*
   - The mass can be accessed with a standard L5 laminectomy.
   - Bone may be removed from the pedicles and facets to facilitate exposure in some cases, but that is unlikely to be necessary here.
   - The dura should be opened with preservation of the underlying arachnoid to prevent rapid cerebrospinal fluid (CSF) release, which may obscure the operative field, increase the amount of epidural bleeding, and increase the risk of neurologic injury.
   - The dural edges are tacked back and the arachnoid is opened sharply and clipped to the dura with vascular clips.
   - The expected intraoperative findings will vary with the tumor type.
   - Meningiomas commonly arise lateral or posterior to the neural elements, which are usually pushed aside.[2]
     - Invasion of neural structures is very rare.
     - If there is no risk of neurologic injury from doing so, the tumor should be removed en bloc to prevent spillage of neoplastic cells.
     - The dural attachment is variably extensive and should be coagulated thoroughly or excised if possible.
   - Schwannomas tend to arise from dorsal nerve roots and come from a single fascicle that can be seen entering and exiting the tumor mass.
     - Surrounding fascicles associated with the tumor's pseudocapsule can often be dissected free.
     - If not, the tumor may be debulked, leaving the pseudocapsule and adherent fascicles behind.
   - In contrast, neurofibromas often arise from ventral nerve roots and may involve and invade multiple adjacent fascicles.[1,2]
     - The involved nerve root is usually nonfunctional and frequently must be sacrificed to achieve a gross total resection.
   - Myxopapillary ependymomas generally arise from the conus area[5] and are surrounded by lumbosacral nerve roots, requiring piecemeal removal.
6. *What operative adjuncts might you consider?*
   - Intraoperative ultrasound can be used to determine the limits of the tumor prior to dural opening.
   - An operating microscope should always be used.
   - Direct electrical stimulation of nerve roots with bipolar stimulation and electromyography may be used to identify important functional roots.
   - Electrophysiological monitoring with somatosensory evoked potentials (sensitivity 87%, specificity 90%) and motor evoked potentials (MEPs) (sensitivity 100%, specificity 75%) can be used to detect potentially reversible neurologic injuries.[1]
   - A cavitating ultrasonic aspirator can be used to help debulk the interior of the tumor.
7. *What are the potential complications of surgery in this case, and what can you do during surgery to reduce the risk of complications?*
   - The risk of neurologic morbidity is 15%.
   - The risk of neurologic worsening is minimized by avoiding manipulation of the nerve roots, spinal cord, and other neural elements and by utilizing aggressive bony removal when necessary.
   - The CSF spaces should be cleared of blood by thorough irrigation prior to dural closure to reduce the risk of arachnoiditis and aseptic meningitis.
   - A watertight dural closure using dural patch grafts when necessary will decrease the risk of CSF leak and pseudomeningocele formation.
   - The risk of spinal instability in cases requiring extensive bone removal can be reduced by concurrent instrumentation and fusion.[1]

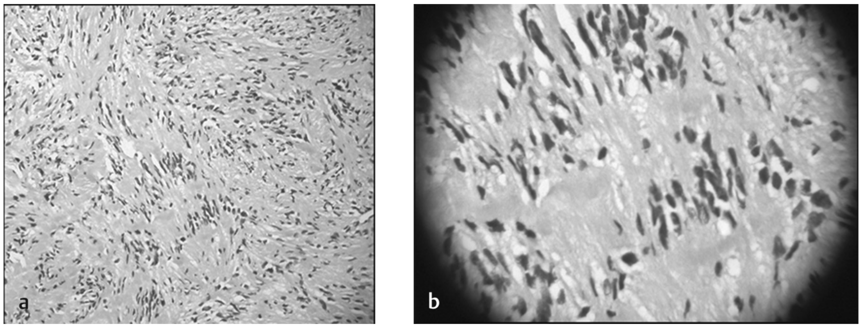

**Fig. 127.2** Hematoxylin and eosin stains of pathology specimen: representative (**a**) low- and (**b**) high-power views are shown.

## Answers (*continued*)

8. *Hematoxylin and eosin stains of the specimen at low and high power are shown (▶**Fig. 127.2**). Describe the histopathology. What is the diagnosis?*
   - Multiple spindle-shaped cells with tapering nuclei are seen arranged in compactly associated parallel streams. This is typical of the Antoni A pattern.
   - More loosely textured Antoni B areas are also visible, primarily in the upper left corner of the image.
   - Verocay bodies are apparent in the higher magnification image.
   - These features are all consistent with the diagnosis of schwannoma.[5]
9. *Describe the cytogenetics associated with this tumor.*
   - Alterations of chromosome 22q, including loss of the entire chromosome, loss of heterozygosity (LOH) of 22q, or loss of function mutations of the *NF2* gene found at 22q, are the only consistent genetic alterations found in schwannomas.[6]
10. *What is the expected outcome of surgery in this case, and what is the potential for recurrence?*
    - Significant relief of pain and improvement of neurologic symptoms are seen in 80 to 90% of cases.[1,7]
    - Schwannomas have a 6 to 12% recurrence rate with a mean time to recurrence of 5 years.
    - Subtotal resections have a recurrence rate of less than 15%.[1,2]

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# Case 128 Intramedullary Spinal Tumor

*Christopher D. Baggott and Amgad S. Hanna*

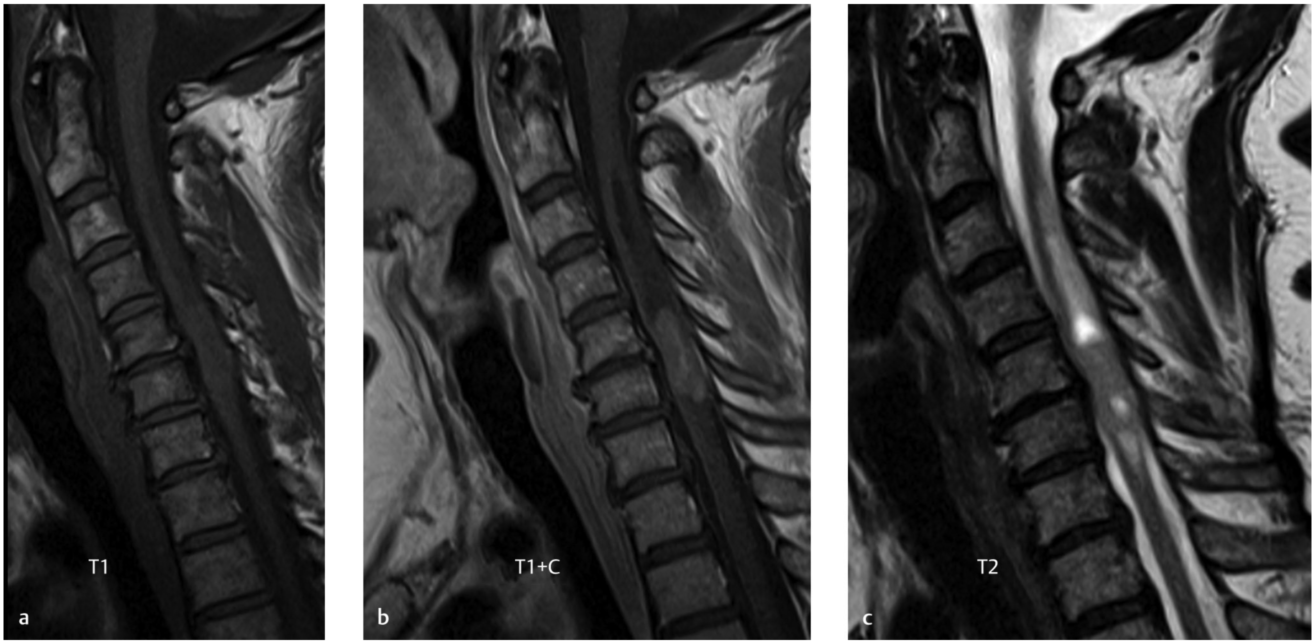

**Fig. 128.1** This sagittal MRI of the C-spine reveals an intramedullary mass with associated syrinx. The mass is isointense to the spinal cord on T1 images. There is heterogeneous enhancement after gadolinium administration. On T2-weighted images, the mass is isointense to the spinal cord, with a hyperintense syrinx extending rostrally and caudally.

## Clinical Presentation

- A 52-year-old woman presents with a 6-year history of symmetric tingling in the arms and legs. Recently, this progressed to numbness in the left arm/hand and loss of dexterity of both hands. She has noticed worsening balance, but no loss of bowel or bladder function.
- Neurologic examination shows normal cranial nerves, no papilledema, symmetric strength, 2+ reflexes, bilateral Hoffman's sign, left ankle clonus, an upgoing toe on the left to plantar stimulation, and patchy loss of light touch and pin prick in all extremities.

## Questions

1. What is the clinical differential diagnosis?
2. What tests can be ordered to narrow the differential diagnosis?
3. Describe the imaging findings in ▶**Fig. 128.1**. What is the differential diagnosis based on this patient's imaging?
4. What are some helpful imaging findings on MRI to determine the most likely pathology?
5. What indications would you use to recommend surgery?
6. What is the goal of surgery?
7. What is the role of intraoperative neurological monitoring (IOM)? How does this affect the anesthetic choice?
8. What is your expectation for the patient's postoperative neurologic condition?
9. The pathology is shown in ▶**Fig. 128.2**a. What is your diagnosis?
10. The postoperative MRI demonstrates gross total resection. What is her prognosis? Does she need adjuvant treatment?

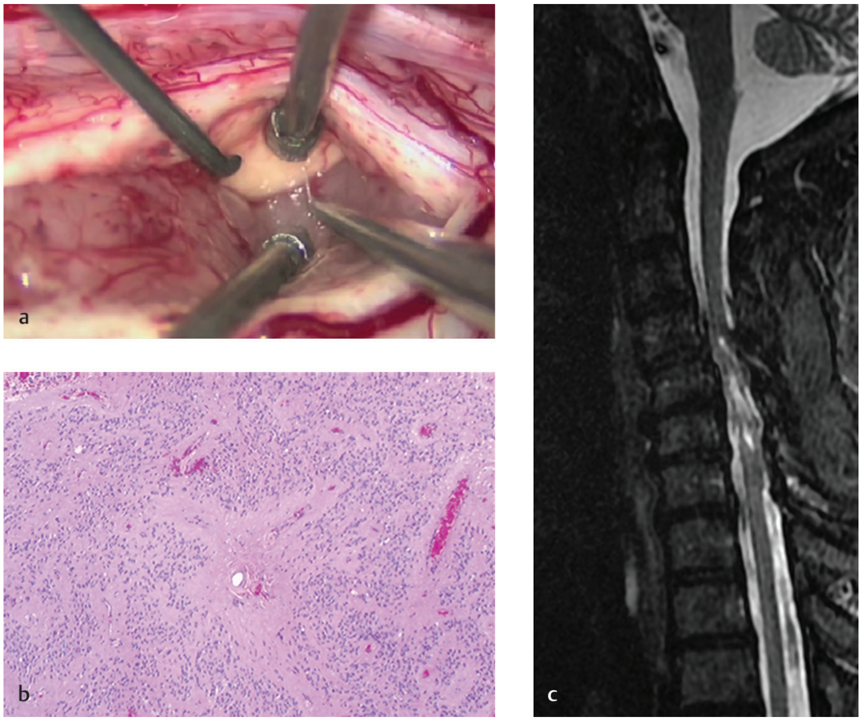

**Fig. 128.2** (**a**) Magnified operative view using the microscope. Note the plane evident between the gray tumor and the surrounding spinal cord. (**b**) Histological section revealing perivascular pseudorosettes, characteristic of ependymoma. (**c**) Postoperative T2-weighted MRI reveals no tumor.

## Answers

1. *What is the clinical differential diagnosis?*
   The differential diagnosis based on history and examination should start with the neuroanatomic localization of the lesion. Based on the anatomic localization, the etiological differential diagnosis should be considered.
   - Anatomic localization
     – The sensory symptoms involving the upper and lower extremities localize the lesion to the cervical spinal cord, the brainstem, the peripheral nerves, or bilateral cerebral hemispheres. The absence of headache and facial symptoms, the absence of papilledema, and the rarity of bilateral symmetric cerebral lesions makes it less likely to be cerebral pathology. The absence of cranial nerve involvement makes it less likely to be brainstem pathology. The development of pathologic reflexes such as the Hoffman's sign makes it less likely to be peripheral nerve pathology. This is most likely a cervical spinal cord lesion.
   - Etiological differential diagnosis[1,2]
     – *Degenerative*: disc herniation, cervical stenosis, instability or deformity; degenerative pathologies are usually associated with neck pain, radiculopathy, and/or myelopathy.
     – *Neoplastic*: extradural (metastasis, primary bone tumor), intradural extramedullary (meningioma, schwannoma, neurofibroma, ganglioneuroma), or intramedullary (ependymoma, astrocytoma, hemangioblastoma). The chronic progressive symptoms in this case fit with a benign neoplastic etiology.
     – *Vascular*: Cavernous malformation and arteriovenous malformations (AVMs); AVMs usually present with sudden or stepwise decline while cavernous malformations can have a more indolent course. Spinal cord infarcts present acutely, usually with motor symptoms due to involvement of the anterior spinal artery.
     – *Demyelinating*: multiple sclerosis (MS). The progressive course fits with an MS plaque, but often symptoms are relapsing and remitting with MS; the age of onset in MS is usually younger. Transverse myelitis and neuromyelitis optica (NMO) typically present acutely or subacutely with more dramatic deficits. Transverse myelitis is generally a short segment of the spinal cord, while NMO is generally a longer segment of the spinal cord (≥ four levels)
     – *Trauma*: central cord syndrome, acute spinal cord injury, spinal cord injury from cervical spine instability
     – *Toxic/metabolic*: alcoholism, vitamin B12 deficiency (subacute combined degeneration)
     – *Developmental*: syringomyelia from a Chiari malformation or tethered spinal cord, enteric cysts, dural ectasia, and arachnoid cysts
     – *Neurodegenerative*: leukodystrophies, motor neuron disease (amyotrophic lateral sclerosis and spinal muscle atrophy), muscular dystrophies,

## Answers (*continued*)

and peripheral nerve disorders (Charcot–Marie–Tooth disease)
  - *Others*: sarcoidosis, tuberculosis, lymphoma, fungal or viral infections, radiation myelopathy, epidural abscess, spontaneous epidural or subdural hematomas (usually associated with anticoagulation), and acquired immunodeficiency syndrome (AIDS) are less likely from the history.

2. *What tests can be ordered to narrow the differential diagnosis?*
   The most important test to order to narrow the differential diagnosis is a cervical spine MRI with and without gadolinium.
   - With a syrinx in the absence of a mass lesion, order a total neuraxis MRI to look for cranial or tethered cord pathologies. Myelomalacia may warrant evaluation for cervical instability with flexion-extension X-rays. Dilated vasculature or sudden neurologic changes may warrant evaluation with spinal angiography.
   - Laboratory work-up to aid in narrowing the differential diagnosis if a spinal cord tumor is not found. This might include vitamin B12 level, methyl malonic acid and homocysteine levels, serum and urine protein electrophoresis (SPEP and UPEP), cerebrospinal fluid (CSF) evaluation for oligoclonal bands (MS) or NMO antibodies, CSF or serum evaluation for angiotensin-converting enzyme (ACE; sarcoidosis); Quantiferon gold (tuberculosis); CSF fungal/viral titers based on exposure history.
3. *Describe the imaging findings in* ▶ **Fig. 128.1**. *What is the differential diagnosis based on this patient's imaging?*
   Imaging demonstrates a spinal cord expanding mass lesion at the C5–C6 levels with an associated syrinx from C3 to C7. There is patchy enhancement on T1 with contrast. The mass lesion is isointense on both T1 and T2. The syrinx is hypointense on T1 and hyperintense on T2. The differential diagnosis includes ependymoma (most common in this age group), astrocytoma (second most common), hemangioblastoma, cavernous malformation, and metastasis.[3]
4. *What are some helpful imaging findings on MRI to determine the most likely pathology?*
   - Imaging findings[1,2,3]:
     - *Ependymoma*: generally centrally located on axial imaging, often with a well-defined border and patchy contrast enhancement; there is commonly a T2 dark "cap sign" from hemosiderin at the poles of the tumor associated with ependymomas.
     - *Astrocytoma*: often eccentrically located on axial imaging with a poorly defined border, "cap sign" is uncommon.
     - *Hemangioblastoma*: a large syrinx with a small, avidly contrast-enhancing mural nodule is characteristic; hemosiderin T2 hypointensities can be seen; often subpial in location.
     - *Cavernous malformations*: usually a "popcorn-like" appearance on T2, irregular but well-circumscribed borders; associated with hemosiderin T2 hypointensities; often subpial on the anterolateral aspect of the spinal cord in location.
5. *What indications would you use to recommend surgery?*
   Indications for surgery include the following[4]:
   - Tissue diagnosis
     - Thorough laboratory evaluation including serum and CSF should be completed prior to spinal cord biopsy for lesions that are not obviously neoplastic but are increasing in size.
     - Intraoperative pathology should be obtained with essentially all surgeries to rule out nontumor pathology and guide resectability.
   - Neurologic deficits
     - Neurologic deficits incurred before surgery are unlikely to improve after surgery, so preoperative neurologic condition predicts functional status postoperatively. If observation is chosen, new neurologic deficits are unlikely to recover.
     - Lesions that are subpial (cavernous malformations and hemangioblastomas) and more central lesions (ependymomas and astrocytomas) that are accessible by a posterior midline myelotomy approach are surgically accessible with reasonable morbidity.
   - Asymptomatic hemangioblastomas and cavernous malformations can often be followed radiographically, treating the lesions if they grow or become symptomatic.
   - Risk/benefit of surgery should be weighed carefully.
     - Risks include neurologic deficits such as paraplegia or quadriplegia, loss of bowel or bladder function, respiratory failure, tracheostomy and ventilator dependency, CSF leak, neck pain, cervical instability, tumor recurrence, hematoma, infection, and spinal cord ischemia.
     - Benefits include reducing the risk of progressive neurologic decline and diagnosis for the appropriate use of adjuvant treatment as necessary.
6. *What is the goal of surgery?*
   The primary goals of surgery are diagnosis and decompression of the spinal cord.
   - If it is safe, a gross total resection is the goal for ependymoma and cavernous malformation, both of which often have a distinct border.
   - Astroyctomas often have an indistinct border, making maximal safe resection the best treatment, while often followed radiographically; adjuvant radiation (or chemotherapy if the tumor is high-grade and has a favorable molecular marker profile) is sometimes used.

## Answers (*continued*)

- Hemangioblastomas should be treated with opening the cyst and resection of the mural nodule; there is no need to resect the wall of the cyst.

7. *What is the role of intraoperative neurological monitoring (IOM)? How does this affect the anesthetic choice?*
   - IOM generally includes somatosensory evoke potentials (SSEP) and motor evoked potentials (MEP). Generally, the loss of SSEP is of little importance, but preservation of SSEP signals allows the surgeon to proceed with aggressive intervention. MEP are more closely associated with postoperative motor function.[4] A 50% decrease in MEP amplitude should be a warning sign to the surgeon of impending neurologic injury. IOM generally requires total intravenous anesthesia (TIVA) without chemical paralysis.
8. *What is your expectation for the patient's postoperative neurologic condition?*
   - Most often, patients are slightly worse neurologically after surgery. Severe deficits from surgery usually do not recover, but minor deficits tend to improve with time. Long-term postoperative neurologic status reflects preoperative neurologic status, making it imperative to operate on patients before severe neurologic deficits occur. While in the immediate postoperative period, diffuse hyperesthesia and paresthesias are common for several days; these resolve in most patients, although severe persistent sensory disturbances can occur. Proprioception deficit is often encountered with approaches adjacent to the dorsal columns. This improves over 6 to 12 weeks in most patients. Motor function improves over a longer period of time, generally seeing improvement in deficits for about 1 year after surgery. Late neurologic deterioration implies tumor recurrence until proven otherwise.
9. *The pathology is shown in* ▶ **Fig. 128.2***a. What is your diagnosis?*
   - ▶ **Fig. 128.2**a demonstrates characteristic features of an ependymoma.[5] This hematoxylin and eosin (H&E) stained section demonstrates a monomorphic hypercelluar field with a fibrillary background. The nuclei are round and regular. Many cells are arranged in perivascular pseudorosettes with long processes extending from the nuclei to the wall of blood vessels, creating a perivascular nuclear-free zone, giving the rosette appearance. True rosettes, where the processes for a central channel without a blood vessel at the center, are mainly seen in medulloblastomas, but can be seen in ependymomas. Alternatively, astrocytomas would have more pink cytoplasm adjacent to a nucleus that has a salt-and-pepper chromatin pattern. Rosettes and pseudorosettes would not be seen. Hemangioblastomas would have pleomorphic cells in a dense background of staghorn blood vessels with abundant lipid droplets in the cytoplasm. Cavernous malformations would have back-to-back thin-walled blood vessels without intervening central nervous system (CNS) parenchyma.
10. *The postoperative MRI demonstrates gross total resection. What is her prognosis? Does she need adjuvant treatment?*
    - Postoperative T2-weighted MRI reveals no residual tumor. If this is confirmed by gadolinium-enhanced MRI, the prognosis is very good because complete resection of an ependymoma can be curative. Clinical as well as radiologic observation should be recommended at this point. Radiation therapy should be recommended only in cases of incomplete resection.[4–8]

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# Case 129 Spinal Metastases

*Christopher Evan Stewart, Brian Seaman, and Joseph A. Shehadi*

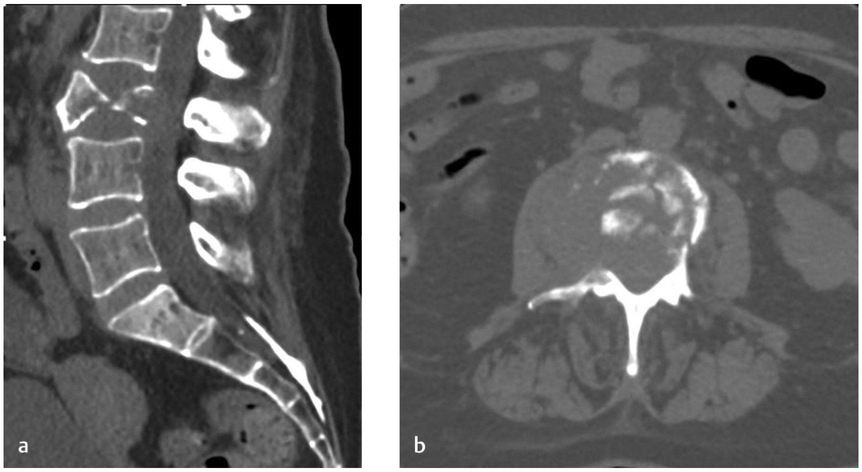

**Fig. 129.1** CT lumbar spine sagittal view and axial view through the L3 vertebral body.

## Clinical Presentation

- A 62-year-old, otherwise healthy female with no prior medical history presents with 4 months of progressively worsening low back pain after moving some boxes. Her complaints also include sharp radiating pain to the anterolateral thighs, most pronounced in the left leg, in addition to some numbness and tingling around both knees. Her pain had become so severe that she could not navigate her own home.
- On neurologic examination, she had Glasgow Coma Score (GCS) 15 and cranial nerves II to XII were intact. On gait testing, she was nonambulatory. She was able to stand but not in the erect position. Muscle strength was 5/5 in all muscle groups of the bilateral lower extremities, supine exam. She had minor sensory abnormalities in the areas superior and medial to her bilateral knees. Deep tendon reflexes were diffusely hypoactive. Physical exam was also positive for cervical lymphadenopathy and an irregular, nonpigmented scalp lesion.
- Her postvoid residual volumes were greater than 200 cc, consistent with urinary retention.
- A CT scan of the lumbar spine was performed and the images are shown in (▶**Fig. 129.1**).

## Questions

1. Interpret the CT scan of the lumbar spine (▶**Fig. 129.1**).
2. What is your initial management for this patient? What additional studies would you order?
3. Interpret the MRI of the lumbar spine (▶**Fig. 129.2**).
4. List a broad differential diagnosis for extradural spinal lesions.
5. List the most common tumors to metastasize to the spinal column in the adult population.

   CT of the chest, abdomen, and pelvis did not reveal any evidence of visceral primary or metastatic disease. MRI of the head revealed a subcutaneous 1.6 × 1.3 cm enhancing mass which correlated to her scalp lesion.
6. In a patient with spinal metastases, what novel and comprehensive frameworks are available to direct decision-making?
7. What is the patient's score using the Spinal Instability Neoplastic Score (SINS) classification. What does this imply?
8. Select a surgical treatment strategy for this lesion.

   The patient acutely underwent surgical intervention. The goals of surgery were to obtain tissue diagnosis, prevent neurologic decline, alleviate pain, and stabilize the affected motion segments. The patient underwent staged anterior and posterior procedures. Through a retroperitoneal approach, an L3 corpectomy was performed with

## Questions (*continued*)

resection of the tumor burden along with arthrodesis and placement of an expandable cage and anterolateral plating of L2–L4. This was followed by posterolateral arthrodesis and pedicle screw fixation from L2 to L5. The following week she underwent a complete excision of the scalp lesion. Histopathologic evaluation of the L3 lesion and scalp lesion confirmed the diagnosis of malignant melanoma. After healing of her surgical wounds, she began two-agent targeted chemotherapy and radiation therapy. At her 6-week follow-up, she had regained ability to ambulate and had minimal pain. Unfortunately, at her 12-week follow-up, her oncologist had found radiographic evidence of diffuse metastatic disease to her lung, spleen, liver, and pelvis.

9. What spinal tumors tend to be radiosensitive? What is the typical fractionation schedule for individuals with spinal metastasis?
10. What are the benefits of surgical decompression in addition to radiation compared with radiation therapy alone?
11. Is there a role for percutaneous vertebroplasty and kyphoplasty in metastatic disease?
12. What complications must you consider in individuals who are considered for surgical intervention who have previously been treated with radiation therapy?

## Answers

1. *Interpret the CT scan of the lumbar spine (▶* **Fig. 129.1***).*
   - CT scan shows a compression fracture of L3 with > 50% loss of height. There is no evidence of malalignment.
   - The vertebral body has a lytic appearance.
   - There is evidence of soft tissue burden within the spinal canal.
   - The CT findings are consistent with a pathologic fracture.
2. *What is your initial management for this patient? What additional studies would you order?*
   - With insidious onset of symptoms and CT findings suggesting a pathologic fracture, an oncologic etiology must be considered. Given the CT results and neurologic symptomatology, an MRI with and without contrast of the areas in question should be performed.
   - In a patient with urinary retention and evidence of spinal canal compromise, a Foley catheter should be placed.
   - Corticosteroids are indicated for the prevention of neurologic deterioration and analgesia.[1] In addition, there may be an oncolytic effect for certain histology types (e.g., breast cancer and lymphoma).[2] Intravenous (IV) dexamethasone should be initiated if the patient does not have contraindications to corticotherapy. Standard dosing comprises a bolus of 8 to 10 mg IV followed by 4 mg IV every 6 hours. Due to gastrointestinal side effects, H2 blockers or proton pump inhibitors are recommended.
   - A CT scan of the chest, abdomen, and pelvis with IV contrast should be performed to search for a primary lesion, or for restaging if a known primary exists.
   - A total body bone scan, using technetium-99m (Tc99m)-labeled phosphate compounds, can be used to determine whether a lesion is solitary or multifocal. Uptake depends on local blood flow and osteoblastic activity; therefore, malignancies such as multiple myeloma are not usually detected via this method.
3. *Interpret the MRI of the lumbar spine (▶* **Fig. 129.2***).*
   - T2-weighted sagittal and axial images demonstrate an isointense lesion at L3 with significant epidural extension and resultant stenosis. T1-weighted sagittal and axial images with gadolinium injection demonstrate the involvement of the vertebral body, bilateral pedicles, and Batson's plexus.
   - There is near complete loss of the cerebrospinal fluid (CSF) space.
4. *List a broad differential diagnosis for extradural spinal lesions.*
   - Metastasis
   - Benign or malignant primary spinal tumors, such as chordoma
   - Retroperitoneal sarcomas
   - Infectious lesions such as osteomyelitis and Pott's disease
5. *List the most common tumors to metastasize to the spinal column in the adult population.*
   - More than 90% of spinal tumors are metastatic.
   - Breast, lung, and prostate cancer make up 60 to 70% of neoplasms that metastasize to the spinal column and cause symptomatic spinal cord compression.[3,4]
   - Less frequently, lymphoma, renal cell carcinoma, colon carcinoma, melanoma, and sarcomas metastasize to the spinal column.[2,5]
6. *In a patient with spinal metastases, what novel and comprehensive frameworks are available to direct decision-making?*
   - The decision of palliative versus aggressive treatment of spinal metastases has traditionally been guided according to the estimated patient survival. The prior prognostic scoring systems included the modified Tokuhashi Scoring System and Tomita scores. These have been criticized because they do

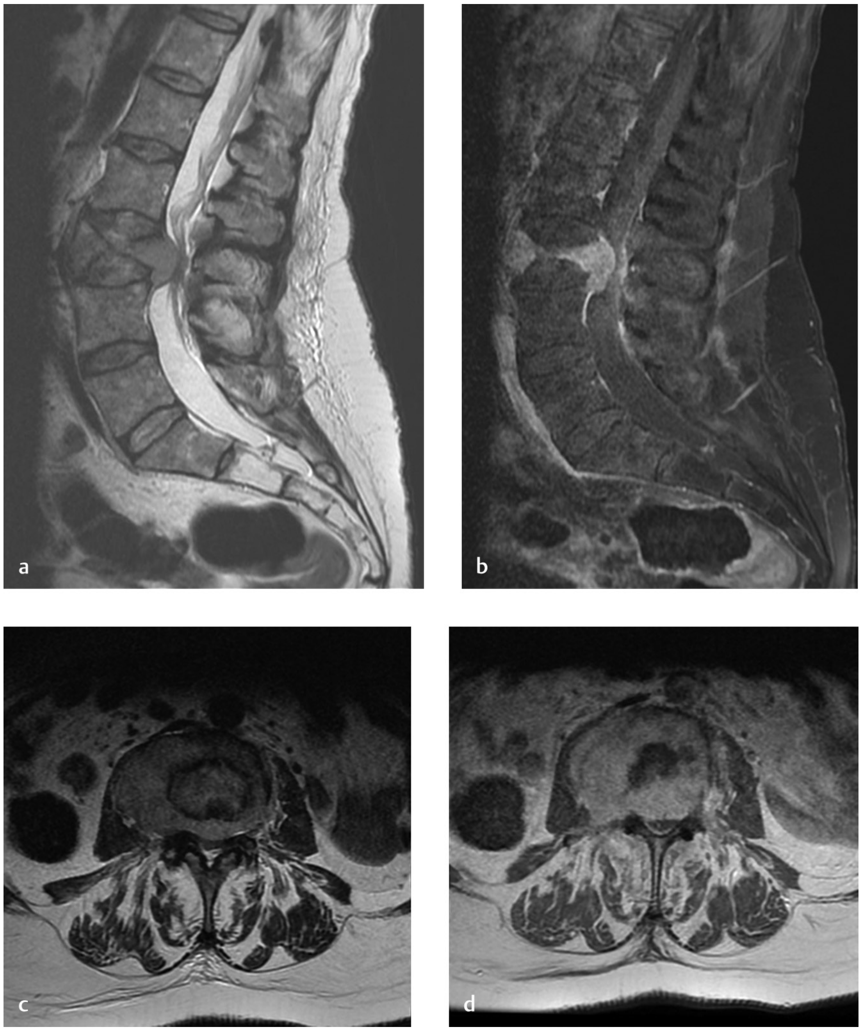

**Fig. 129.2** (**a–d**) MRI of the lumbar spine including T2-weighted sagittal and axial views as well as gadolinium-enhanced T1-weighted sagittal and axial views.

## Answers (*continued*)

not consider the presence of instability or technological advancements in radiosurgery.[6]

- Recently a novel decision-making framework focusing on four fundamental assessments has been proposed. The NOMS framework focuses on the **N**eurologic, **O**ncologic, **M**echanical, and **S**ystemic considerations in a patient with spinal metastases to determine the optimal radiation and surgical treatments.[6,7]

7. *What is the patient's score using the Spinal Instability Neoplastic Score (SINS) classification. What does this imply?*
   - The patient's SINS score is 13. This consists of 2 points for location, 3 points for pain, 2 points for lytic bony lesion, 0 points for alignment, 3 points for > 50% loss of height, and 3 points for bilateral posterior element involvement.
   - The score of 13 delineates this as an unstable lesion.[8]
8. *Select a surgical treatment strategy for this lesion.*
   - This patient is otherwise healthy, but does have significant pain-mediated disability, and neurologic impairment (radiculopathy and urinary retention). This is combined with an isolated spinal lesion with an unstable SINS score of 13, and lack of a tissue diagnosis.
   - In this particular scenario, a biopsy and noninvasive oncologic treatment would be insufficient. Surgical decompression and stabilization followed by tissue diagnosis and appropriate radiotherapy and chemotherapy is a good option.
   - The lesion is unstable and vertebral body incompetent. Isolated laminectomy for decompression should not be considered as it fails to address the site on neural compression and further destabilizes the spine.
   - With a large ventral tumor burden and posterior element involvement, a combined anterior decompression/reconstruction and posterior lateral fusion should be considered.
9. *What spinal tumors tend to be radiosensitive? What is the typical fractionation schedule for individuals with spinal metastasis?*

## Answers (*continued*)

- Radiosensitive tumors include breast, prostate, and small cell lung cancer, lymphoma, and multiple myeloma.
- Renal cell carcinoma, melanoma, and non-small cell lung cancer (NSCLC) are relatively radioresistant.[1]
- Typically, total radiation dose ranges from 2500 to 3600 cGy. It is usually given in 10 to 15 fractions to avoid significant side effects.[9]

10. *What are the benefits of surgical decompression in addition to radiation compared with radiation therapy alone?*
    - A randomized trial[10] assigned 101 patients with cord compressions to either radiotherapy alone ($n = 51$) or decompression surgery followed by radiotherapy ($n = 50$).
    - Results demonstrated that surgery plus radiotherapy is superior to radiotherapy alone in the treatment of spinal cord compression caused by metastasis.
    - Patients who underwent surgery were able to walk more than 3.5 times longer than those who received radiotherapy alone: a median of 126 days versus 35 days, respectively ($p = 0.006$).
    - Of the 16 patients in each group who entered the trial and were unable to walk, 56% of those in the surgery arm regained mobility, compared with 19% of patients who received radiation alone ($p = 0.03$).
    - Overall survival was not affected.[9]
11. *Is there a role for percutaneous vertebroplasty and kyphoplasty in metastatic disease?*
    - Percutaneous vertebroplasty and kyphoplasty represent a successful option for the palliative treatment of intractable spinal pain associated with malignant spinal tumors and metastases.[11]
    - In a randomized controlled trial, pain and quality of life have shown to be improved in cancer patients with compression fractures treated with balloon kyphoplasty.[12]
12. *What complications must you consider in individuals who are considered for surgical intervention who have previously been treated with radiation therapy?*
    - Spinal radiation before surgical decompression for metastatic spinal cord compression is associated with a significantly higher wound complication rate.[13]
    - Risk of wound infection may be up to three-fold higher than in those individuals undergoing surgery de novo.[9]

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# Case 130 Lumbar Vertebral Mass

*Achal Patel, Danial Branch, and Juan Ortega-Barnett*

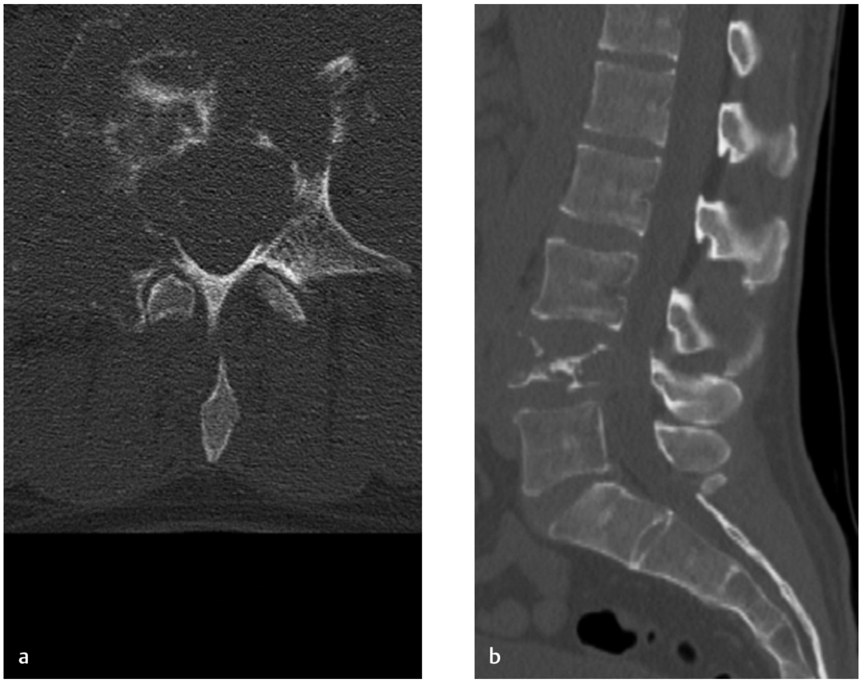

**Fig. 130.1** (**a, b**) CT scan without contrast shows expansile lytic lesion in the L4 vertebral body.

## Clinical Presentation

- A 39-year-old male presents with low back pain and left leg radicular pain in an L4 distribution for 1 month.
- The back pain is constant with a burning, shooting pain down the left leg.
- He is able to ambulate but has an antalgic gait with significant back pain upon sitting or standing.
- He denies any fevers, chills, weight loss, or recent injury/trauma. Bowel and bladder function are normal.
- Neurological exam revealed numbness in the L4 dermatome in the left leg without weakness. Reflexes were normal. There was no perianal anesthesia. No other neurological deficits were found.
- Noncontrast CT and MRI T1-weighted images with gadolinium are shown in ▶**Fig. 130.1** and ▶**Fig. 130.2**, respectively.

## Questions

1. Describe the findings on the CT and MRI scans (▶**Fig. 130.1** and ▶**Fig. 130.2**).
2. What further work-up would you like?
3. What is the differential diagnosis?
4. Biopsy reveals plasmacytoma. What further tests do you require?
5. What are the treatment options?
6. Describe the anterolateral approach for a corpectomy.
7. What are the risks associated with doing a corpectomy?
8. Describe the outcome for treatment of plasmacytoma.

The surgery is uneventful, and the patient is transferred to the ICU. Four hours later, the nurse calls you concerned that after the last wound check, there was what looked like new bruising on the patient's right flank. You immediately go see the patient, who is now complaining of inguinal area pain. On examination, you appreciate that the patient is alert and oriented, but hypotensive and mildly tachycardic. The wound is well approximated and there is no oozing, but you do confirm the flank bruising.

9. What are the next steps in management and what could be occurring?

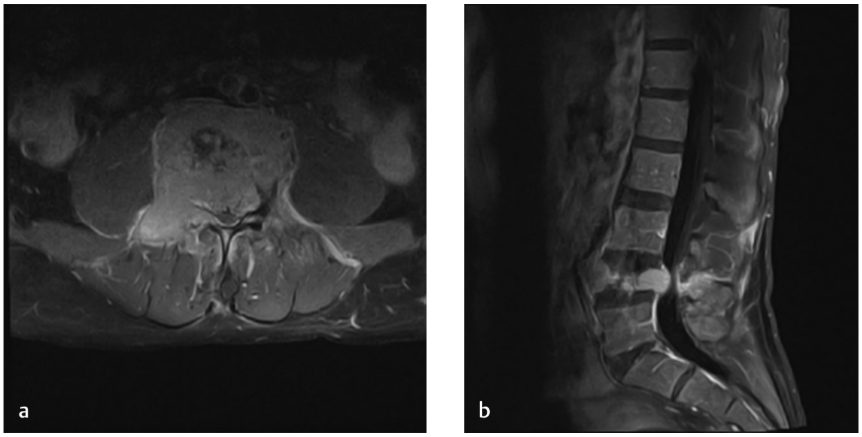

**Fig. 130.2** (**a, b**) MRI with gadolinium shows enhancement of lesion causing compression of thecal sac.

## Answers

1. *Describe the findings on the CT and MRI scans (▶* **Fig. 130.1** *and ▶* **Fig. 130.2***).*
   - CT scan shows a lytic and expansile lesion involving all three columns of the L4 vertebrae.
   - MRI reveals homogenous enhancement of the vertebral body lesion with severe central canal stenosis and compression of the thecal sac.
2. *What further work-up would you like?*
   - Complete blood count, chemistry panel, liver function panel (aspartate aminotransferase [AST], alanine aminotransferase [ALT]), prothrombin time (PT) and partial thromboplastin time (PTT)
   - CT chest/abdomen/pelvis
   - Serum and urine protein electrophoresis, prostate-specific antigen (PSA), C-reactive protein
   - Consider positron emission tomography (PET) CT scan of the whole body and CT-guided biopsy
3. *What is the differential diagnosis?*
   - Multiple myeloma, plasmacytoma, metastasis, giant cell tumor, chordoma, chondrosarcoma, osteoblastoma, osteochondrosarcoma
   - Less likely infections such as osteomyelitis and epidural abscess
4. *Biopsy reveals plasmacytoma. What further tests do you require?*
   - Further studies include skeletal survey and bone marrow biopsy.
   - True diagnosis of solitary plasmacytoma of the spine requires the following: negative skeletal survey except for the solitary spine lesion, biopsy diagnosis, and no evidence of plasma cell dissemination, satisfied by normal calcium, creatinine, and hemoglobin levels and no plasmacytosis on bone marrow biopsy.[1]
5. *What are the treatment options?*
   - Treatment plan for solitary plasmacytoma of the spine can vary to some extent. These malignant tumors respond well to radiation, and that is often the primary treatment.
   - However, surgical intervention is warranted for spinal instability and/or decompression of the neural elements and sometimes needed for tumors resistant to radiotherapy.[2] Surgical resection may also improve local control and progression-free survival, and possibly decrease the radiation dose needed.[1]
   - Adjuvant radiation treatment should also be considered, even in cases of en-bloc resection.[3] This combined treatment may delay time to local recurrence and the likelihood of developing multiple myeloma in the future.[4,5]
   - Chemotherapy may increase disease-free survival[6] but its role for solitary plasmacytoma is unclear.
   - If there is no sign of spinal instability, then only radiation is needed. However, in the setting of spinal instability, surgical intervention involving instrumentation is warranted and easier to perform prior to radiation to avoid radiation-related surgical complications.
   - In this case, the entire vertebral body, both pedicles, right facet joint, and bilateral lamina are infiltrated by tumor. Using the Spinal Instability Neoplastic Score (SINS),[7] a total of 14 points is calculated which suggests instability (3 points for location in mobile spine L2–L4, 3 points for pain with movement, 2 points for lytic lesion, 0 points for normal alignment, 3 points for greater than 50% collapse of the vertebral body, and 3 points for involvement of the bilateral posterolateral elements). Therefore, surgical intervention for stabilization of the spine is needed for this patient prior to radiation.
   - Posterior instrumentation from L3 to L5 using pedicle screws and rods via a percutaneous or open approach is one option. Additional L4 corpectomy with expandable cage placement can be done for correction of significant kyphosis > 20 degrees. This would relieve the anterior compression of the thecal sac, and increase the rigid fixation of the construct. Furthermore, corpectomy or complete spondylectomy reduces the tumor burden and

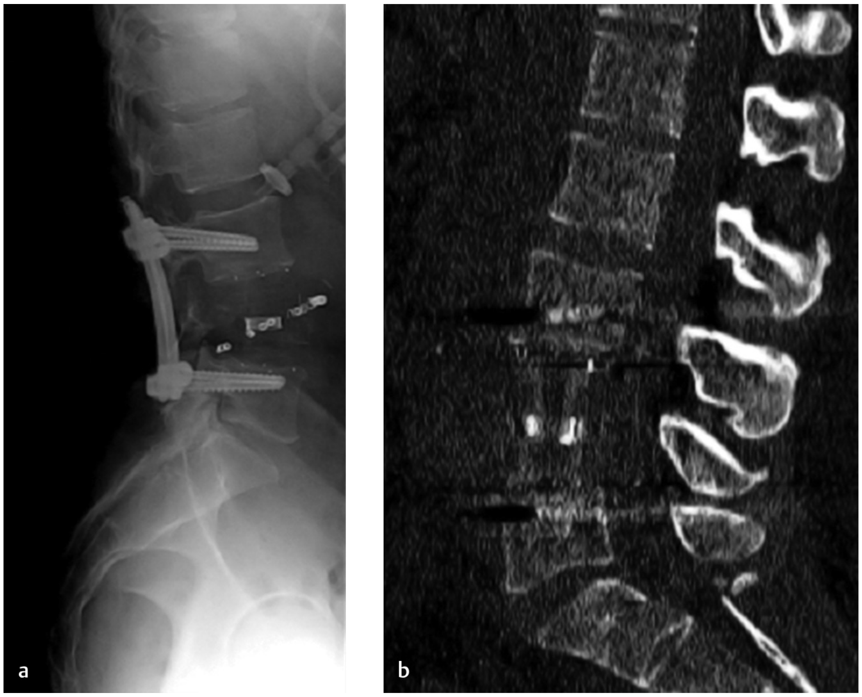

**Fig. 130.3** Plain X-ray films (**a**) and CT scan (**b**) shows anterior corpectomy with cage placement and posterior instrumentation.

## Answers (*continued*)

volume; although it is not clear to what extent volume reduction prior to radiation is needed to significantly prevent recurrence and increase survival. However, in the absence of malalignment, posterior instrumentation with or without laminectomy for decompression can be considered in light of a patient's comorbidities and it also limits surgical blood loss, even if there is significant collapse of the vertebral body secondary to tumor. Complete spondylectomy with anterior and posterior instrumentation via separate anterior and posterior approaches or through a single posterior approach is another option.[8]

- Preoperative embolization has been suggested to reduce blood loss when doing a spondylectomy as these tumors are often highly vascularized.[9]
- Radiotherapy should be done about 6 weeks after surgical fusion to allow for osteoblast differentiation.[10]
- In this case, an anterior approach for corpectomy with expandable cage placement and minimally invasive posterior instrumentation was performed, as shown in ▶**Fig. 130.3.**

6. *Describe the anterolateral approach for a corpectomy.*
    - The patient should be positioned in the semilateral position. One then performs a diagonal incision on the patient's left side from the posterior aspect of the 12th rib to the lateral border of the rectus abdominis, making sure that it overlaps the level of interest. Working on the left side allows for easier repair of the aorta, if accidentally injured, and it is also more resistant to injury than the inferior vena cava.
    - One incises the external and internal oblique muscles and the transversus abdominis muscles to expose the retroperitoneal fat. The fat is then bluntly dissected to expose the psoas muscle.
    - The retroperitoneum is then retracted medially, along with the ureter. The aorta and the common iliac arteries and veins should also be retracted medially. Segmental lumbar arteries off the aorta can safely be tied off if they obstruct the operative view of the level of interest.
    - Needle localization within the disc space can then be used to identify the correct level.
7. *What are the risks associated with doing a corpectomy?*
    - Risks include injury to the nearby vascular structures, such as the aorta, the inferior vena cava, and the radicular arteries, retrograde ejaculation secondary to injury to the superior hypogastric plexus, tearing of the ureter, injury to the laterally lying sympathetic chain, and injury to the genitofemoral nerve overlying the psoas muscle.
    - It is thus essential to identify these aforementioned structures and manipulate them with care.
    - Other risks associated with spine surgery in general include instability, nonunion, pseudoarthrosis, cerebrospinal fluid (CSF) leak or fistula, direct nerve injury or nerve damage (thecal sac, nerve root, or plexus), resulting in weakness, numbness, or bowel and bladder problems, infection, hematoma. In particular, in this case, there is also risk of tumor residual and/or recurrence.

## Answers (*continued*)

- General risks of surgery can also include deep venous thrombosis, pulmonary embolisms, myocardial infarction, and death.

8. *Describe the outcome for treatment of plasmacytoma.*
   - Median survival is 10 years with radiation treatment.[11] Fifty percent of cases will progress to multiple myeloma.[11]
   - Some studies have suggested better prognosis with surgical curettage and adjuvant radiation, but the degree of tumor burden reduction that translates into a significant change in outcome following radiation is unclear.
   - Poor prognostic factors include tumor size[12] and persistence of M protein spike after radiation treatment.[11]
9. *What are the next steps in management and what could be occurring?*
   - The patient is complaining of groin pain and clinically has a Grey Turner's sign. He is also hypotensive. Retroperitoneal hematoma, a rare but recognized complication of the surgery, must be ruled out.
   - Initial management should focus on the ABCs, and fluid resuscitation should begin. Bloodwork should be ordered immediately, including complete blood count, chemistry panel, PT, PTT, international normalized ratio (INR), and renal function tests.
   - Once hemodynamically stable, the patient should be escorted to Radiology for a CT of the abdomen and pelvis. This will enable the diagnosis of the potential hematoma.
   - Should one be found, there is no clear consensus on management strategies. Small hematomas can be managed conservatively, ensuring adequate fluid resuscitation, correction of any coagulopathy, and blood transfusions as needed. Larger hematomas may require surgical evacuation and a General Surgery/Vascular consultation should be sought immediately. Of course, conservative management is only indicated if the patient is entirely hemodynamically stable.[13]
   - Less urgent entities on the differential include injury to the genitofemoral nerve and habitual postoperative pain. Bruising can also occur secondary to positioning, highlighting the importance of protecting all prominent surfaces with bean bags and/or gel pads.

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# Case 131 Cervical Spine Mass

*Juan Ortega-Barnett*

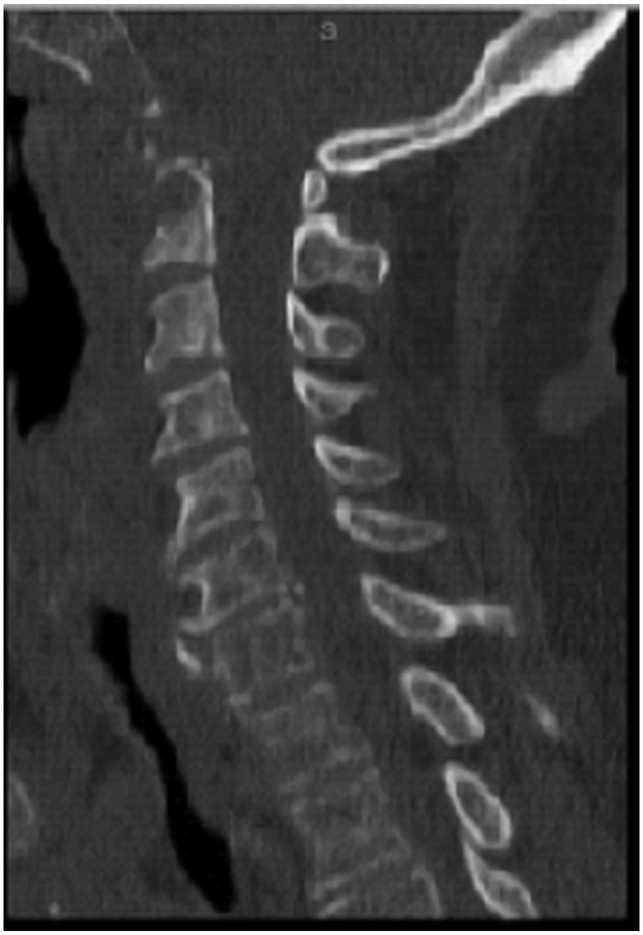

**Fig. 131.1** CT scan sagittal reconstructed view.

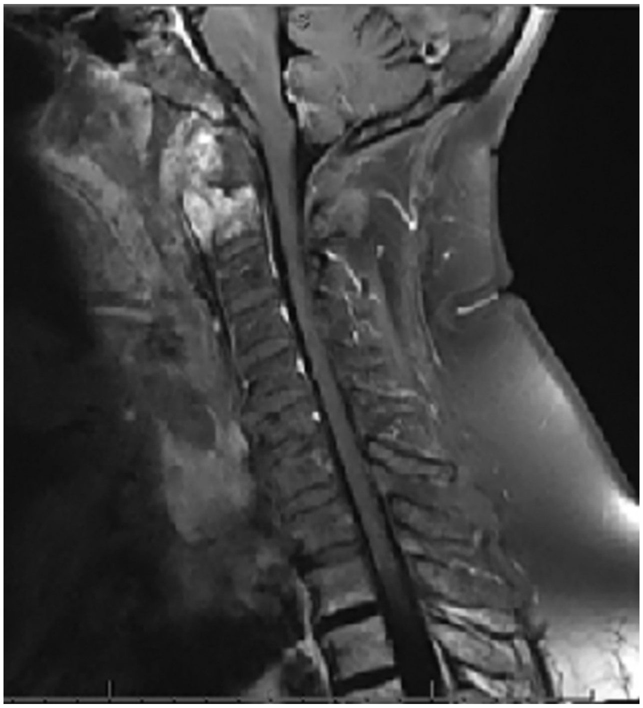

**Fig. 131.2** MRI T1-weighted image with contrast, sagittal view.

## Clinical Presentation

- A 74-year-old African American woman presents with several months' history of chronic neck pain. Five days prior to her admission she woke up with acute and severe neck pain. Examination findings reveal a baseline diabetic peripheral neuropathy and motor assessment is nonfocal.
- Her past medical history is significant for diabetes, hypertension, gout, arthritis, congestive heart failure, coronary artery disease, hyperlipidemia, obesity, interstitial lung disease, and asthma. The patient is a 47-year ppd smoker. Surgical history reveals that she is 7 years post left nephrectomy for renal cell carcinoma T1a, N0, M0, 16 months post right hemicolectomy for adenocarcinoma T2, N0, M0, and 10 months post right breast lumpectomy for hyalinized intraductal papilloma and hyalinized fibroadenoma.
- See ▶**Fig. 131.1** and ▶**Fig. 131.2** for CT and MRI scans.

## Questions

1. Describe the scans.
2. What further studies would you order?
3. What is the differential diagnosis for this lesion?
4. What are the common clinical manifestations of these lesions?
5. What is your initial management for this patient?
6. Which are the most common tumors to metastasize to the spinal column?
7. Which metastatic tumors are radiosensitive and which are radioresistant?
8. What definitive management would you offer to this patient?
9. What are specific factors and potential surgical pitfalls to consider given the pathological diagnosis in this particular case?

## Answers

1. *Describe the scans.*
   - CT scan sagittal view of the craniocervical junction shows severe bony erosion of C1 anterior arch and C2 odontoid process with associated soft-tissue mass.
   - There is a pathologic dens-impacted fracture and lateral luxation of the C1 lateral masses. Craniocervical junction involvement with possible stenosis is suspected on the CT scan. Severe damage of C1, C2, and the atlantoaxial articulation is observed.
   - There is also degenerative disc–osteophyte complex disease at C5–C6 and C6–C7.
   - MRI scan T1 sagittal view with contrast shows enhancement with bone destruction of C1, C2, and the lower third of the clivus.
   - MRI of the cervical spine and the craniocervical junction also shows an anterior vertebral mass centered in the prevertebral space and resulting in lytic destruction of the anterior aspects of C1 and C2, and the inferior aspect of the clivus.
   - No spinal canal stenosis or spinal cord signal abnormalities are identified.
2. *What further studies would you order?*
   A full metastatic and medical work-up should be completed including:
   - CT scan of chest, abdomen, and pelvis
   - Positron emission tomography (PET) CT scan of the entire body
   - Mammogram
   - Basic laboratory profile: complete blood count (CBC), electrolytes, prothrombin time/partial thromboplastin time (PT/PTT), type and screen
   - Tumor marker serum levels such as: carcinoembryonic antigen (CEA) or cancer antigen (CA)-125
   - Oncology consultation to evaluate the cancer status of the patient
   - Medical clearance including: cardiology and pulmonary clearance with referral to the appropriate specialists and testing, such as electrocardiography (EKG), pulmonary function tests, echocardiogram if indicated, and other cardiopulmonary blood markers

   CT scans of chest, abdomen and pelvis were also obtained here. In this case, the study identified two left subpleural nonspecific nodules.
3. *What is the differential diagnosis for this lesion?*
   - Several possible etiologies of spinal column lesions would include infectious, inflammatory, or neoplastic.
   - Infections to consider: vertebral osteomyelitis that may be pyogenic or nonpyogenic. Nonpyogenic etiologies would include: tuberculous spondylitis, brucellosis, aspergillosis, blastomycosis, coccidioidomycosis, and infection with candida.[1,2]
   - Lesions associated with neurosarcoidosis with osseous involvement occur in up to 13% of patients, but skull and vertebral involvement in neurosarcoidosis is very rare. The intervertebral body disc spaces can also be affected. Lesions are usually lytic with a punched-out appearance, but can also be sclerotic. The osseous lesions can also show increased activity on bone scans.[3]
   - Primary spinal bone column lesions include chordomas, chondrosarcoma, osteoblastoma, osteochondroma, vertebral hemangioma, aneurysmal bone cyst (ABC), giant cell tumors, osteogenic sarcoma, brown tumor of hyperparathyroidism, and giant cell granuloma (variant of ABC).
   - Other tumors include plasmacytoma, multiple myeloma, chloroma, eosinophilic granuloma, some cases of Ewing's sarcoma can be primary of the spine (rare).[4,5]
   - Spinal metastasis account for the majority of the epidural and bone tumors, and is the most likely diagnosis in this case given the medical history and appearance of the lesion on imaging studies. Metastasis to the C1–C2 region comprise only around 0.5% of all spinal metastases.[6]
   - This patient did not have any fever, had a normal white blood cell count and sedimentation rate. She does have a very significant history for previous primary cancer treatments. Her CT and MRI results point to spinal metastasis as a more likely diagnosis.

## Answers (*continued*)

4. *What are the common clinical manifestations of these lesions?*
   - These C1–C2 spinal column tumors will present with suboccipital and posterior cervical pain. Occasionally, patients find themselves grabbing their head in order to support its weight and minimize the pain when they are in an upright position. However, it is characteristic that the patient with spinal metastasis will experience increased pain when recumbent, especially at night.
   - In the case of spinal metastasis to the cervical region, between 11 and 15% of patients present with neurologic symptoms. However, 15% of these patients develop cord compression and around 6% have quadriplegia.[6]
   - In patients with rheumatoid arthritis with cervical spine involvement, up to 25% were found to have atlantoaxial subluxation that manifest with local pain in 67% of cases, with spasticity and paresis in nearly 27% of cases, and with sensory disturbance in 20%.[7]
5. *What is your initial management for this patient?*
   - Initially cervical stabilization with external orthotics should be implemented, such as a C-collar, Minerva brace, or a halo.
   - The patient should be admitted to the hospital. Corticosteroids are indicated for the prevention of neurologic deterioration and analgesia. Gastrointestinal (GI) prophylaxis and pain control.
   - Once CT chest, abdomen, and pelvis have been completed, a planned CT-guided biopsy for tissue diagnosis might be considered.
   - However with this patient, because of her possible neck instability and amount of pain, the decision was made to perform an open biopsy under general anesthesia at the same time as planned surgical stabilization of her cervical spine.
   - The patient should receive medical, cardiac, and pulmonary clearance prior to any type of surgical intervention given her extensive medical history.
6. *Which are the most common tumors to metastasize to the spinal column?*
   - Common primary sites of tumors that metastasize to the spinal column include lung, breast, prostate, kidney (renal cell), lymphoma, GI tract, melanoma, multiple myeloma, and thyroid cancer.[8]
7. *Which metastatic tumors are radiosensitive and which are radioresistant?*
   - Tumors that do not respond well to radiation therapy include renal cell carcinoma, melanoma, and non-small cell lung carcinoma. Those that have a better response are prostate, breast, small cell lung cancers, lymphoma, and multiple myeloma.[9]
8. *What definitive management would you offer to this patient?*
   - In certain cases, some authors would offer chemotherapy and radiation with simultaneous external immobilization of the spine. Some osteoblastic tumors with pathologic fractures, such as prostate metastases or some breast metastases, may heal with radiation treatment and immobilization alone.[10]
   - Anterior approaches for stabilization at this location are difficult. A lateral approach, such as the one used to access foramen magnum tumors, is well described for spinal tumors in this location.[11,12]
   - However, in this case, because of the extension of the tumor into the clivus as well as the first two cervical vertebrae, her extensive medical comorbidities, and prognosis, a decision was made to perform an occipital cervical fusion down to the C5 level. Biopsy confirmed renal cell carcinoma. Her pain was better controlled, hence improving her quality of life. She was managed postoperatively with chemotherapy and radiation therapy.
   - See ▶ **Fig. 131.3** for a postoperative CT reconstruction.
9. *What are specific factors and potential surgical pitfalls to consider given the pathological diagnosis in this particular case?*
   - Renal cell carcinoma metastases are notorious for being very hemorrhagic tumors. This should be considered during the resection and provisions for substantial blood loss should be taken care of such as utilization of cell saver technology in conjunction with the surgical suction in order to be able to provide the patient with autologous transfusion of red blood cells, having banked blood (4 units of packed red blood cells ideally) prepared and cross-matched to the patient's blood type prior to the start of the procedure.
   - If at all possible or if biopsy can be performed preoperatively, then a strong consideration should be given to preoperative angiography with embolization.
   - Adequate hemostasis materials should also be available intraoperatively, such as Gelfoam (Pfeizer) with thrombin, Oxycel (Becton Dickinson), Floseal (Baxter), or other elements such as Avitene (Davol). Finally, renal cell carcinoma should also be studied to its relative (or lack thereof) responsiveness to chemotherapy and radiation therapy. This should be taken into account when devising a postoperative adjunct treatment plan and consideration to additional alternative treatments should also be given.[13,14]

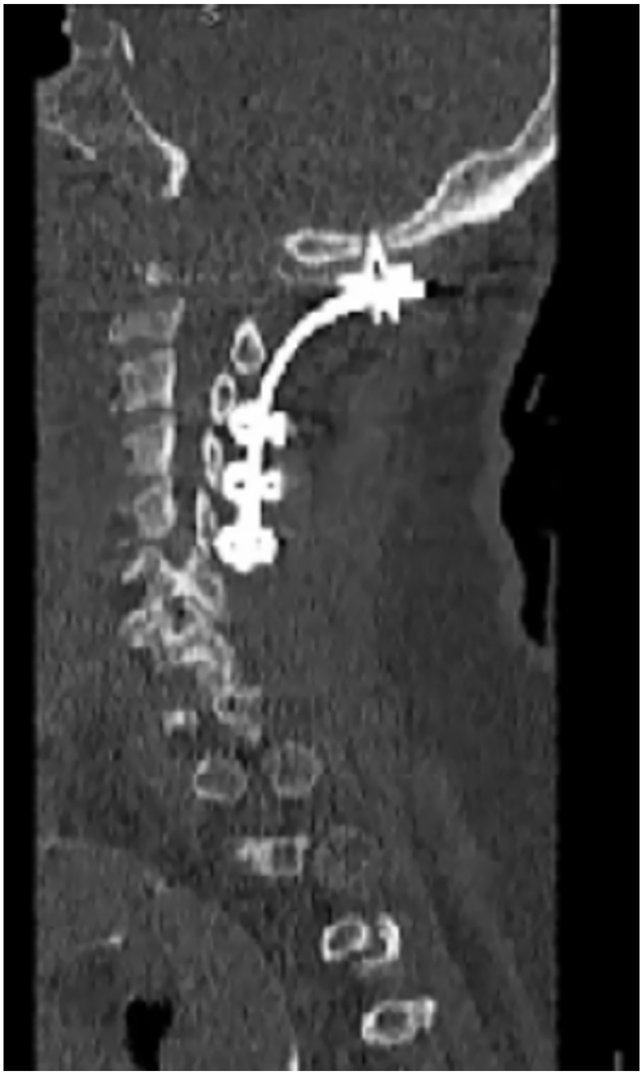

**Fig. 131.3** CT scan sagittal reconstructed view. Postoperative scan demonstrating occipital cervical fusion down to C5.

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# Case 132 Spinal Arteriovenous Malformation

*Bassem Yousef Sheikh*

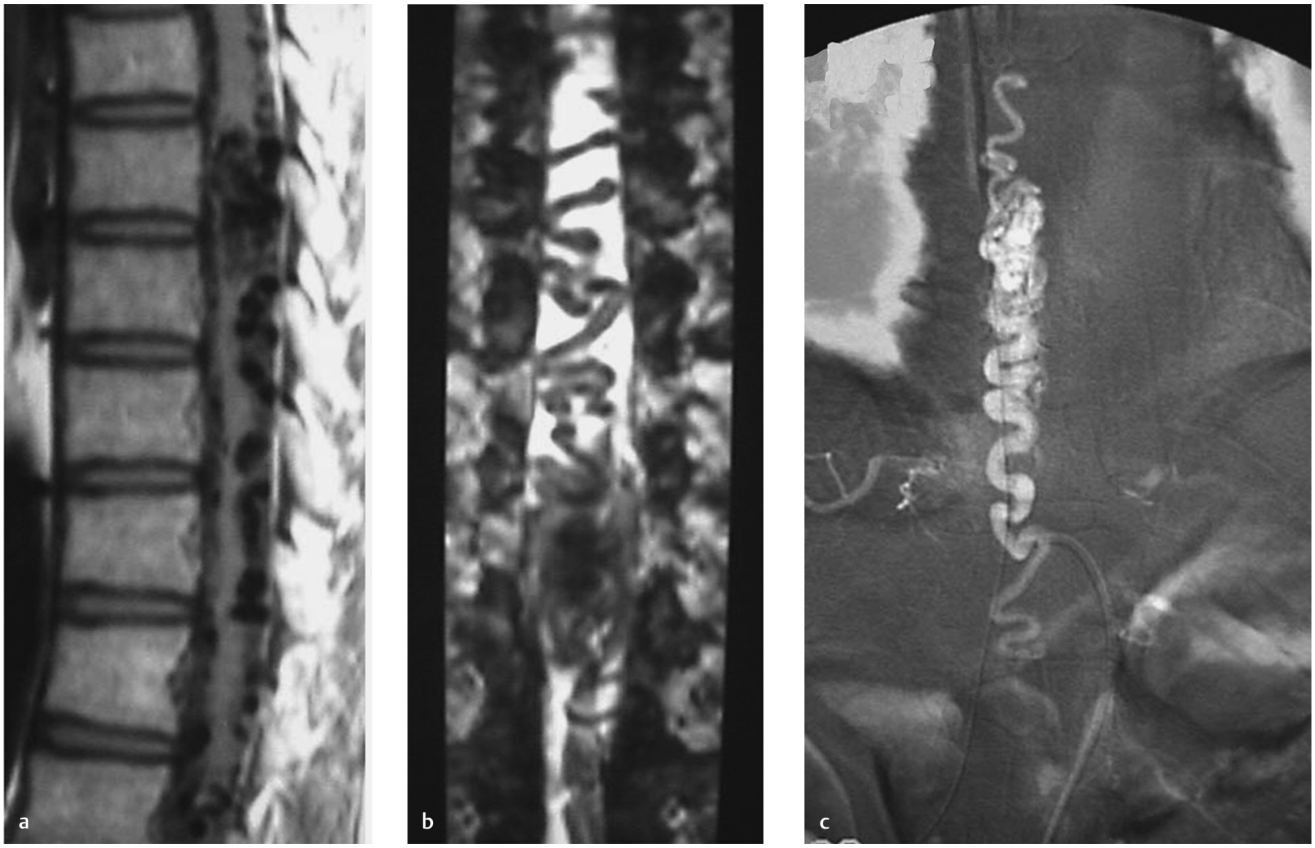

**Fig. 132.1** (**a**) T1-weighted and (**b**) myelographic MRI, and (**c**) selective spinal digital subtraction angiography.

## Clinical Presentation

- A 32-year-old man presents with a 2-year history of progressive paraparesis.
- He has weakness involving both lower limbs that is progressively increasing.
- The patient also complains of back pain and bilateral leg numbness.
- Disturbance of bladder function started 3 weeks prior to presentation.
- He also shows loss of superficial sensation below the umbilicus. The vibration and position sensations are disturbed in both lower extremities.

## Questions

1. Provide a clinical explanation to the patient's presentation.
2. What is your differential diagnosis?
3. Describe a classification of spinal arteriovenous malformations (AVMs).
4. What is your diagnostic work-up for spinal AVMs?
5. Imaging studies are obtained and shown in ▶**Fig. 132.1**. Describe the radiologic findings in these images.
6. What are the therapeutic modalities that may be suggested for this patient?
7. The patient asked you whether he would regain normal neurologic function after treatment. What is your answer to the patient?

## Answers

1. *Provide a clinical explanation to the patient's presentation.*
   - This patient is suffering from a progressive myelopathy.
   - The myelopathy is most likely originating in the thoracic spine area, given the clinical presentation.
2. *What is your differential diagnosis?*
   - The differential diagnosis of spinal progressive myelopathy is broad and includes the following[1,2]:
     - Congenital conditions: Chiari malformation, syringomyelia, narrow canal/short pedicles, mucopolysaccharidoses, kyphosis, os odontoideum
     - Infections: syphilis (infarction), postviral (herpes, varicella, cytomegalovirus), epidural empyema, vertebral osteomyelitis, acquired immunodeficiency syndrome (AIDS)-related myelopathy, tuberculosis (Pott's disease), parasitic cyst
     - Traumatic conditions: spinal shock, epidural hematoma, electrical injury, bone fracture
     - Tumors: spinal cord tumor (extradural, intradural extramedullary, intramedullary), metastases, carcinomatous meningitis, paraneoplastic syndrome
     - Endocrine conditions: Cushing's disease, obesity (epidural lipomatosis), acromegaly, Paget's disease
     - Nutritional conditions or toxins: vitamin B12 deficiency, local anesthetics
     - Degenerative conditions: spondylotic myelopathy, ossified posterior longitudinal ligament, disc herniation
     - Inflammatory or demyelinating conditions: transverse myelitis, multiple sclerosis, Devic's syndrome, Guillain–Barre syndrome, amyotrophic lateral sclerosis
     - Vascular diseases: spinal epidural, subdural, or subarachnoid hematoma, spinal cord infarction (syphilis, aorta clamping intraoperatively, hypotension, aortic dissection), AVM, radiation exposure, reaction to contrast infusion
3. *Describe a classification of spinal arteriovenous malformations (AVMs).*

   There have been several classification systems for spinal vascular malformation in the literature, however, they are broadly categorized into the following three sections[3–6]:
   - Dural arteriovenous fistula (DAVF): This is the most common type of malformation (▶ **Fig. 132.2**).
     - It typically presents in older men.
     - It is usually found in the lumbar and thoracic spine.
     - This lesion consists of a small AVF within or just beneath the dura at the point where a feeding radicular artery enters the dura at the nerve root sleeve (at the level of the intervertebral foramen).
     - The venous outflow from the fistula, carrying arterialized blood, drains into the intradural venous plexus via the radiculomedullary vein along the dorsal surface of the spinal cord.
     - These low-flow fistulae produce venous hypertension (Foix–Alajouanine syndrome) which results in decreased spinal cord perfusion, resulting in intermittent and progressive neurologic deficits.
   - Perimedullary fistula: located intradurally but in the extramedullary space
     - They usually present at the thoracolumbar region.
     - They are characterized by a single shunt without a nidus.
     - They occur between the spinal artery (anterior or posterolateral artery) and the spinal vein.
     - They may ascend rostrally forming craniocervical shunts, reaching even into the posterior cranial fossa.
     - They are located either on the ventral or dorsal surface of the spinal cord.
   - Intramedullary AVMs: These lesions are rare (▶ **Fig. 132.2**).
     - They are supplied by the radiculomedullary artery or the spinal artery.
     - They are found partially or entirely within the substance of the spinal cord.
     - They are further subclassified according to their size and the location within the spinal cord.
       - ○ Large complex metameric AVMs
       - ○ Juvenile type
       - ○ Combined intradural and extradural (intraspinal)
4. *What is your diagnostic work-up for spinal AVMs?*
   - Diagnostic evaluation includes the following:
     - MRI: Visualization of dilated spinal veins is possible, but is difficult in cases of DAVF because the only slightly dilated vessels and the fistula itself cannot be visualized. Increased intramedullary signal in T2-weighted images can reflect the edema due to chronic venous hypertension. Serpiginous areas of low signal owing to signal void in the draining vein may be demonstrated.
     - MR angiography
     - Myelography: Has been replaced by the more informative MRI combined with MR myelographic effect. CT myelography can help define the feeding pedicle location.
     - Selective spinal angiography: This study remains the gold standard of evaluation for spinal AVMs and is the only way of diagnosing and localizing the nidus in dural fistulae. It requires detailed catheterization of thoracic and lumbar arteries.

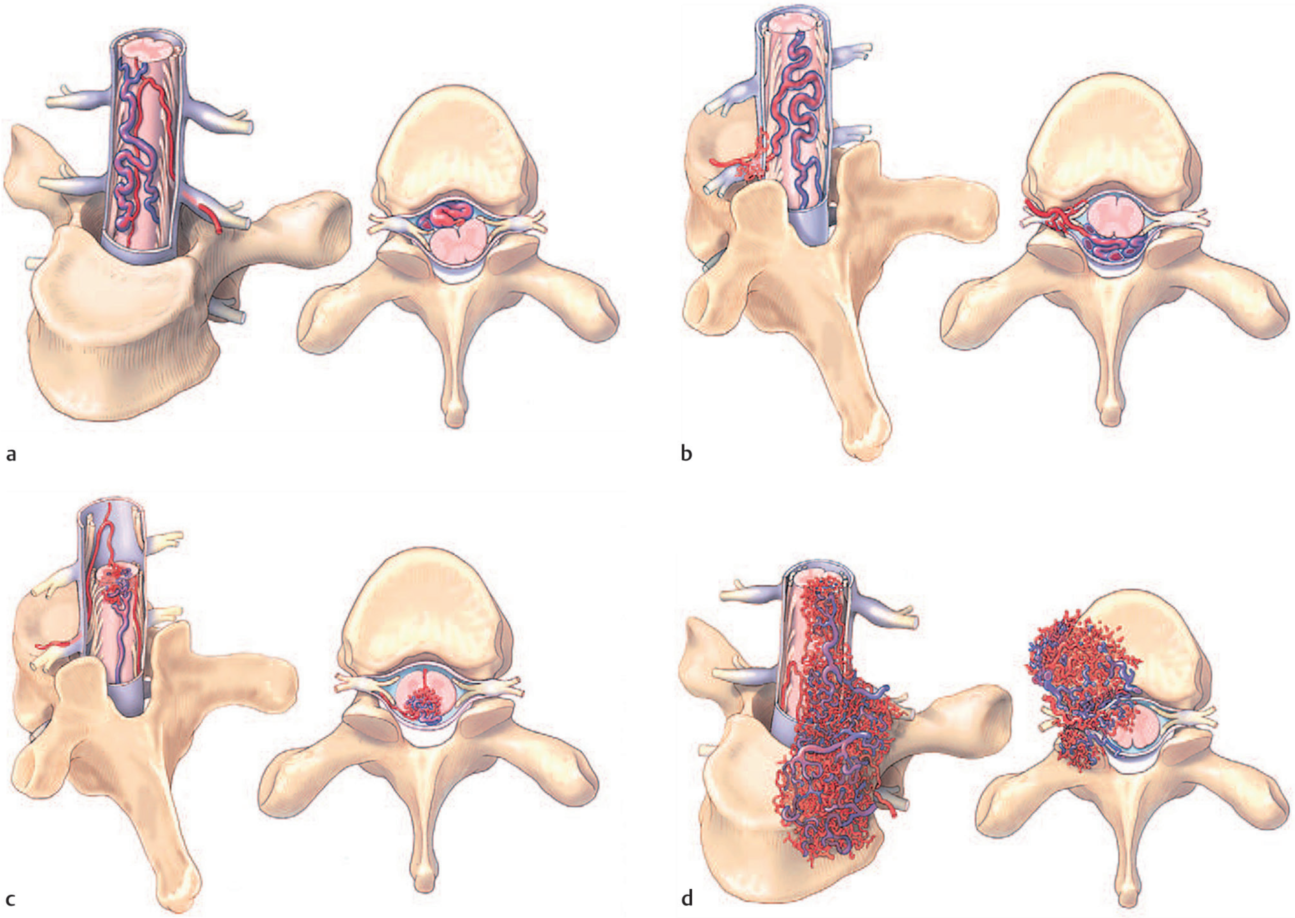

**Fig. 132.2** Classification of spinal arteriovenous malformations (AVMs). (**a**) Anterior and (**b**) dorsal examples of dural arteriovenous fistulae. (**c**) Small compact intranidal spinal AVM and (**d**) large complex metameric spinal AVM. (Reproduced from Gonzalez and Spetzler 2006)[6]

## Answers (*continued*)

If angiographic results are routinely negative, but there is a strong clinical suggestion of a fistula, it may be necessary to extend the investigation to the vertebral, external carotid, or sacral arteries.

5. *Imaging studies are obtained and shown in* ▶**Fig. 132.1**. *Describe the radiologic findings in these images.*
   - These images demonstrate the presence of an intradural, intramedullary AVM that is filling from a spinal artery and draining rostrally and caudally into draining veins.
   - Part of the nidus is present within the spinal cord substance.
6. *What are the therapeutic modalities that may be suggested for this patient?*
   - Successful treatment in each individual spinal vascular malformation requires correct understanding of the lesion's anatomic location and its angioarchitecture.
   - The therapeutic modality that should be suggested depends on the type of spinal vascular malformation.
   - The limitations of both surgery and endovascular embolization should be considered.
   - The following list summarizes each treatment modality based on type of spinal AVM.[3,7,8]
     - DAVF
       - Surgical technique: Surgical exposure to excise the fistula is a simple and successful treatment of spinal DAVFs.
       - Endovascular technique: As an alternative to surgical therapy, injection of *N*-butyl cyanoacrylate can be given after superselective catheterization of the radicular artery. This method has been reported to be successful and definitive in ~75% of cases. If the occlusion of the fistula is attained endovascularly with definitive embolization material, an operation is not necessary. Otherwise, removal of the partially embolized fistula has to be performed.
     - Perimedullary fistulae
       - Surgical elimination: This is an option for posteriorly located accessible small fistulae.

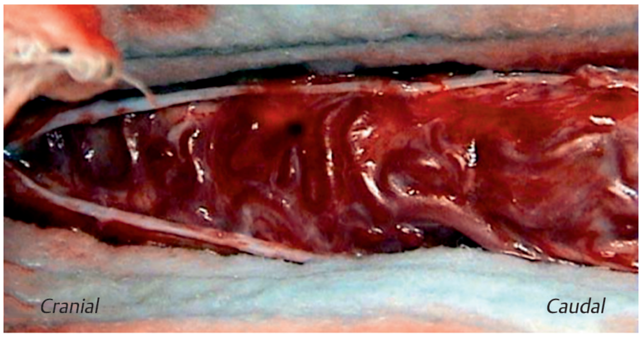

**Fig. 132.3** Intraoperative view of spinal arteriovenous malformation.

## Answers (*continued*)

  - ○ Endovascular embolization: It may be used in the case of ventrally situated fistulae that are inaccessible surgically and giant fistulae with multiple dilated feeders and draining veins.
- – Intramedullary AVMs
  - ○ Endovascular embolization: This is the modality of choice in the management of intramedullary AVMs. Most techniques use particulate embolization materials that result in reduction in the flow through the malformation, thus lowering the steal and ischemic effects on the spinal cord and the venous hypertension. However, particulate embolization is noncurative and revascularization of the malformation should be expected.
  - ○ Surgical technique: Microsurgical therapy alone is sometimes technically difficult owing to the intramedullary and ventral location of the AVM. ▶**Fig. 132.3** illustrates an intraoperative view of such an AVM.

7. *The patient asked you whether he would regain normal neurologic function after treatment. What is your answer to the patient?*
   - Postoperative improvement of patients with neurologic deficits depends on preoperative duration of signs and symptoms, and on the degree of disability.[7,8]
   - Improvements may usually involve both sensory and motor deficits.
   - The genitosphincteric disturbances have a much more severe prognosis and persist more often.
   - Because of progressive ischemic lesions of the spinal cord caused by chronic venous congestion, the shunt should be eliminated as early as possible.[9]

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# Case 133 Spinal Arteriovenous Fistula

*Badih Daou, Pascal M. Jabbour, and Erol Veznedaroglu*

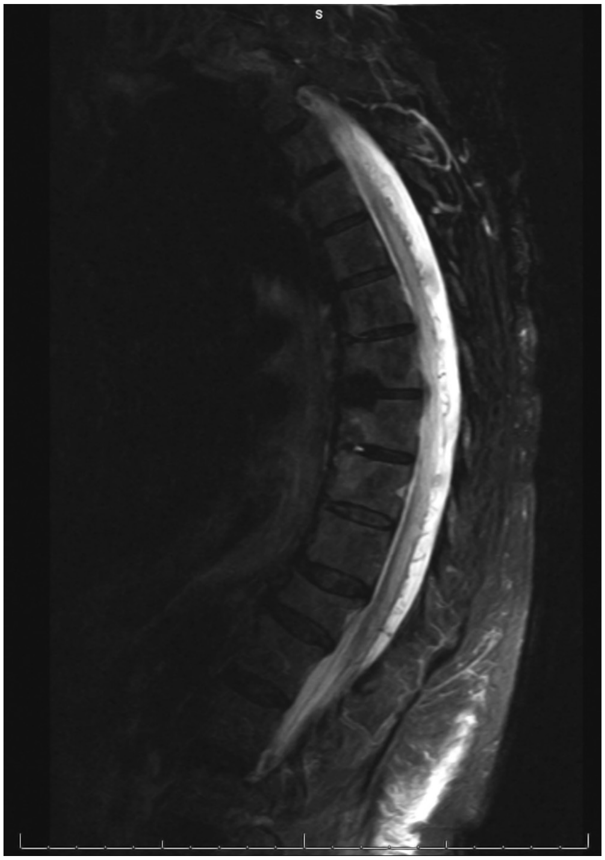

**Fig. 133.1** T2-weighted MRI, midsagittal section.

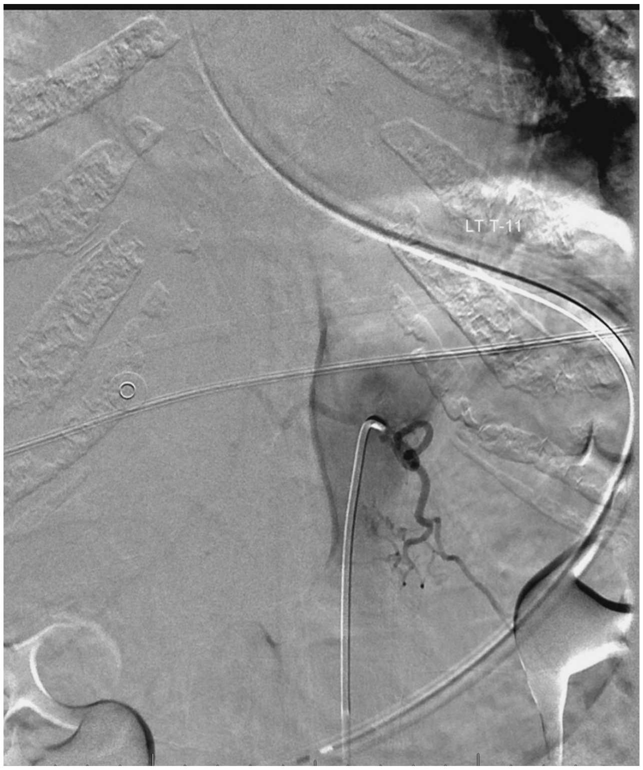

**Fig. 133.2** Thoracic spine angiogram, anteroposterior view with selective injection of thoracic artery.

## Clinical Presentation

- A 77-year-old woman with a history of high blood pressure and diabetes presents to the emergency room for progressive weakness in her lower extremities.
- The weakness has been ongoing for the last 8 months, associated with episodes of bladder incontinence for last 3 weeks.
- She denies any lower back pain or radicular pain. There is no history of trauma.
- Neurologic examination reveals weakness in her proximal lower extremity muscle groups graded at 3/5 with spasticity. Distally, her strength is graded 4/5 on the Medical Research Council scoring system. Sensation is intact. She has increased deep tendon reflexes and a right Babinski sign. Her rectal tone is normal. She has normal strength in the upper extremities.
- MRI of the thoracic spine is shown in ▶**Fig. 133.1.**

## Questions

1. Where do you localize the lesion according to her examination?
2. Describe the MRI and angiogram findings.
3. What is the next imaging test to perform? (▶**Fig. 133.2**)
4. What are some common characteristics of spinal dural arteriovenous fistulas (DAVFs)?
5. What are some clinical differences in spinal DAVFs and spinal intradural arteriovenous malformations (AVMs)?

## Questions (*continued*)

6. What are the different treatment options for this patient?
7. What are some complications of management of spinal DAVFs?
8. What are the outcomes of treatment of spinal DAVFs?

## Answers

1. *Where do you localize the lesion according to her examination?*
    - She has upper motor neuron signs with increased reflexes, Babinski sign, and spasticity. Her upper extremities are normal; therefore, the lesion is probably located at the level of her thoracic spine.
2. *Describe the MRI and angiogram findings.*
    - The MRI shows edema in the spinal cord with prominent flow voids representing engorged vessels surrounding the spinal cord. On MRI, the combination of cord edema, perimedullary dilated vessels, and cord enhancement is characteristic.
    - The angiogram demonstrates a DAVF, which is also a type I spinal AVM.
3. *What is the next imaging test to perform? (▶* **Fig. 133.2***)*
    - A spinal CT angiogram or a spinal formal angiogram should be performed. Spinal angiogram is shown in ▶**Fig. 133.2**.
4. *What are some common characteristics of spinal dural arteriovenous fistulas (DAVFs)?*
    - Spinal DAVFs are the most common type of spinal vascular malformations, accounting for 70% of these malformations.
    - DAVFs are most commonly found in the thoracolumbar region.
    - These fistulas are created when a radiculomeningeal artery feeds directly into a radicular vein, leading to engorgement of radicular veins and the venous system of the spinal cord resulting in intramedullary venous hypertension that may cause compression of the spinal cord or nerve roots.
5. *What are some clinical differences in spinal DAVFs and spinal intradural arteriovenous malformations (AVMs)?*
    - Spinal DAVFs occur predominantly in male patients, whereas intradural AVMs are more equally distributed in both genders.
    - The age of occurrence in spinal DAVFs is older than in intradural AVMs (46 vs. 24 years).
    - Onset of symptoms is more gradual in spinal DAVFs. Clinical symptoms include progressive paraparesis, paresthesias, bladder and bowel disturbances.
    - The first symptom in spinal DAVFs is more likely to be paresis whereas in intradural AVMs it is more commonly related to subarachnoid hemorrhage.
    - Exacerbation of symptoms by activity is more common in spinal DAVFs.
    - The upper extremities are more commonly affected in intradural AVMs.[1]
6. *What are the different treatment options for this patient?*
    - The goals of treatment consist of interrupting the vein draining the AVF as it penetrates the inner dural layer.[2]
    - Her options include:
        - Open surgery with clipping of the fistula that usually resides in the sleeve of a nerve root. Laminectomies one level above and one below are usually performed. The dura can be opened in the midline and retraced laterally. The site of the fistula is identified and correlated with imaging studies. The arterialized vein is then coagulated and a limited resection is performed.[2,3]
        - Endovascular embolization of the fistula:[4,5] Liquid embolic agents including *n*-butyl cyanoacrylate (*n*-BCA) and Onyx (ethylene vinyl alcohol copolymer) are usually used because they are the most likely to fill the distal nidus.
        - Open surgery is preferred if the patient has multiple sites of fistula formation, has tortuous vascular anatomy that may cause difficulty in accessing the arterial feeding vessel, or if the vessel supplies blood to healthy regions of the spinal cord.
        - All procedures should be performed under general anesthesia with neurophysiologic monitoring, depending on the location of the lesion.
7. *What are some complications of management of spinal DAVFs?*
    - Acute neurologic deficit post surgery or post embolization:
        - Paralysis, bladder or bowel dysfunction, or sexual dysfunction from injury to nervous tissue
        - Spinal cord infarction
        - This necessitates immediate imaging studies via MRI, administration of steroids, followed by possible reexploration.
    - Hematoma
    - Postoperative infection
    - Cerebrospinal fluid leak
    - Residual malformation or fistula on postoperative angiography (may require reoperation)
        - It is recommended to perform an angiogram ~1 week after surgical treatment.[2]

## Answers (*continued*)

8. *What are the outcomes of treatment of spinal DAVFs?*
   - In general, safe and successful treatment is achievable.
   - In most cases, there is improvement of neurologic function or treatment—it usually arrests the progression of the symptoms. These good results can be seen in ~80 to 90% of patients.[1,3]
   - Most patients can ambulate after treatment.
   - Bladder symptoms are often improved.
   - Outcome correlates very much with initial neurologic condition upon presentation: patients with less neurologic deficit on presentation will fare better.[1,3] For this reason, prompt diagnosis and treatment play an important role in achieving good outcomes in symptomatic spinal DAVF cases.

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# Case 134 Spinal Epidural Abscess

*Cristion Gragnaniello and Remi Nader*

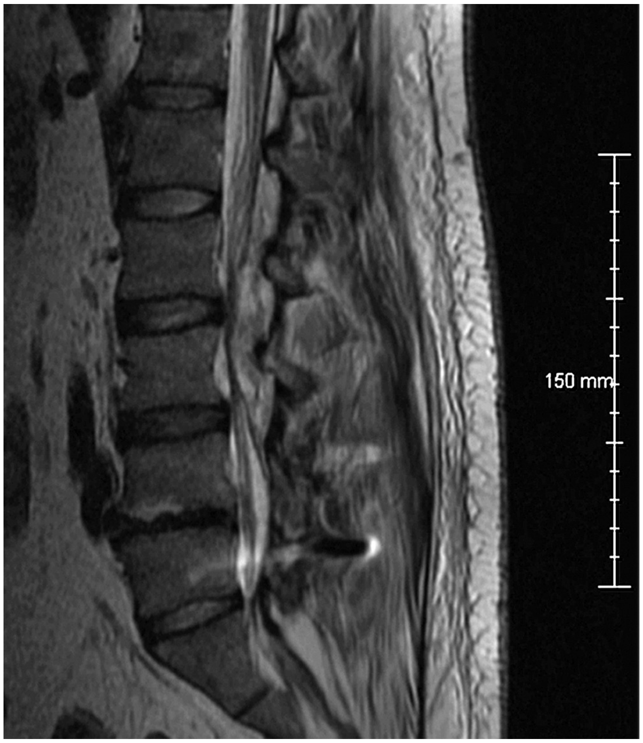

**Fig. 134.1** Sagittal T2-weighted MR image through the lumbar spine.

## Clinical Presentation

- A 39-year-old man presents to your office 2 weeks after he has undergone a hemilaminectomy at L3–L4 for degenerative spinal stenosis.
- He has a previous history of L4–L5 and L5–S1 posterolateral fusion with pedicle screw fixation.
- He presents now with a 2-day history of severe back pain radiating to the buttocks bilaterally.
- There is mild erythema around the recent surgical wound.
- There are no deficits or pain in his lower extremities.
- MRI of the lumbar spine is obtained (▶**Fig. 134.1**).

## Questions

1. Interpret the MRI and give a differential diagnosis.
2. What are the characteristics of epidural abscess on MRI?
3. What are the risk factors for this condition?
4. What are the most common microorganisms involved in epidural abscesses in the immunocompetent patient?
5. What is the incidence of this condition in the post-surgical patient?
6. How do you manage this condition?

You decide to operate. You decompress the thecal sac and perform a sharp debridement of the wound. During surgery, you send cultures and are told that there are numerous Gram-positive cocci. Once down to the thecal sac, you notice that, for the most part, there is thick granulation tissue over the sac. As you tease this tissue off the dura, you get a dural tear.

7. How do you handle a dural tear in this specific setting?

## Questions (*continued*)

8. Why do you not remove the instrumentation of the previous fusion?

After you decompress the thecal sac, you notice a piece of floating bone, you remove it, and you notice that it comprises the inferior facet joint of L3 that is detached and free floating (you did not detach it during your current decompression).

9. Why is the piece of facet detached?
10. What condition do you now worry about, given the detached facet, and how do you manage it?

## Answers

1. *Interpret the MRI and give a differential diagnosis.*
   - There is a posterior epidural collection from L2–L4, hyperintense on T2-weighted images. This could be a cerebrospinal fluid (CSF) collection, abscess, or infected hematoma.[1–4,]
   - The disc space looks normal, and so do the vertebral bodies. This finding is important to assess the radiographic presence of concomitant osteomyelitis.
   - There is loss of lumbar lordosis.
   - Given the clinical presentation, the most likely diagnosis is epidural abscess.
2. *What are the characteristics of epidural abscess on MRI?*
   - On T1-weighted images, it has the same intensity as spinal cord or neural elements.
   - On T2-weighted images, it shows increased signal.
   - On fat-saturated sequences, edema and soft-tissue inflammation are clearly visible and bright.
   - Contrast-enhanced images often show a peripheral rim of enhancement due to granulation tissue and hypervascularity.[1]
3. *What are the risk factors for this condition?*
   - Presence of hardware and instrumentation
   - Repeated surgery
   - Use of steroids
   - Diabetes or other underlying medical problems such as renal failure, previous trauma, and urinary tract infections
   - Intravenous drug abuse[3–5]
4. *What are the most common microorganisms involved in epidural abscesses in the immunocompetent patient?*
   - Gram-positive bacteria such as *Staphylococcus aureus* (reported in 60% of cases) or *S. epidermidis*[4,5]
   - Gram-negative bacteria such as *Escherichia coli* and *Pseudomonas aeruginosa*[4,5]
5. *What is the incidence of this condition in the postsurgical patient?*
   - The generally accepted incidence is less than 10%.
   - The incidence is reportedly increased with higher complexity procedures. It is in the range of 0.6 to 3.7% after microdiscectomy and of 3.7 to 20% after instrumented lumbar cases.[2]
6. *How do you manage this condition?*
   - Look for any discharge so that it can be cultured before starting antibiotics.
   - Explore the wound to see if you find a purulent collection or CSF.
   - Drain the abscess, sampling tissue and purulent material for culture.
   - Perform a sharp debridement and lavage.[3]
   - Place on broad-spectrum intravenous antibiotics until final culture results are available. Vancomycin for the Gram-positive covering including methicillin-resistant Gram-positive cocci and cefepime for Gram-negative bacteria are among the most common medications used.[4]
   - The closure of the wound is very important and can be done in different ways; but the key concept is the obliteration of the dead space caused by the removal of the infected tissue, restoring the blood supply, and postoperative drainage by outflow drain or suction/irrigation device.[6,7]
7. *How do you handle a dural tear in this specific setting?*
   - A tear in this specific condition can lead to a spreading of the infection in the subdural space because of the disruption of the normal dural barrier.[8]
   - Suture with a 7–0 or 8–0 dural suture if accessible in a watertight fashion, using the Valsalva maneuver to confirm the closure.
   - Cover with a dry piece of Gelfoam (Pfizer Pharmaceuticals, New York, NY) or collagen matrix.
   - Cover with fibrin glue or hydrogel sealant.
   - You may need to resect more bone to further the exposure and allow access to the whole tear.[9]
   - Continue appropriate antibiotics.
8. *Why do you not remove the instrumentation of the previous fusion?*
   - The infection on the MRI does not extend to the previously treated levels.
   - The onset of symptoms is very recent as the surgery at the L3–L4 level was performed in the past 2 weeks. Therefore, the formation of the glycocalyx over the rod and other used material is improbable.[2,3]

## Answers (*continued*)

- Titanium materials have a porous surface that allows the penetration of antibiotics.[6]
- Good results have been shown when leaving the instrumentation in place, even in cases where the infection was involving those instrumented levels, if thorough debridement is performed and antibiotic therapy is well managed.[10,11]

9. *Why is the piece of facet detached?*
   - The bone is likely detached due to erosion from osteomyelitis.
10. *What condition do you now worry about, given the detached facet, and how do you manage it?*
    - You worry about two entities:
      - Osteomyelitis and spread of the infection to bone
      - Instability and the possibility of spinal deformity
    - Your management should now involve placing the patient in a brace postoperatively and obtaining flexion-extension lumbar spine radiographs.
    - You may repeat radiographs in 2 to 4 weeks as osteomyelitic changes may become visible radiographically only after 2 to 3 weeks.[3]
    - He may need a lumbar instrumented fusion once the infection has healed if instability is then demonstrated.[12]
    - Treatment with antibiotics does differ in cases of osteomyelitis: he needs 8 to 12 weeks of intravenous antibiotics instead of only 4 to 6 weeks (as in cases of simple epidural abscess with no bony involvement).[5]
    - Hyperbaric oxygenation can help to promote host immune defense response and stimulate vascularization of the injured tissues.[13]

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# Case 135 Vertebral Osteomyelitis and Discitis

*Sohum Desai, Da'Marcus Baymon, and Juan Ortega-Barnett*

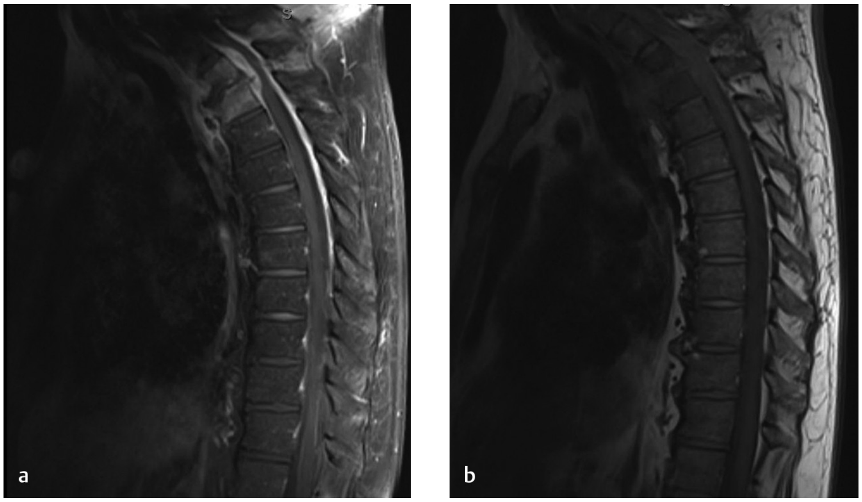

**Fig. 135.1** **(a)** Contrast-enhanced and **(b)** noncontrasted T1-weighted midsagittal thoracic MRI scans.

## Clinical Presentation

- A 68-year-old male presented with 3 months of back pain associated night sweats, hemoptysis, and an unintentional 35-pound weight loss. The day prior to presentation, he reported left lower extremity weakness, anesthesia below the nipples/groin, and urinary and bowel incontinence.
- He has a past medical history of atrial fibrillation, hypertension, and peripheral vascular disease status post stenting. He is being managed with beta-blockers, warfarin, and antihypertensives.
- His physical exam revealed significant point tenderness over the upper thoracic spine, decreased rectal tone, and decreased strength in the left lower extremity. He demonstrated upgoing Babinski bilaterally and hyporeflexia in his patellar and Achilles tendon.
- MRI was obtained and is shown below (▶**Fig. 135.1**).

## Questions

1. What is your differential diagnosis?
2. What initial imaging or lab tests would you order?
3. Interpret the MRI findings.
4. What are the radiographic elements of osteomyelitis/discitis on MRI?
5. What are the risk factors for this condition?
6. What are the common modes of transmission for osteomyelitis/discitis?
7. Describe indications for surgical interventions that may be necessary in a patient who has osteomyelitis/discitis.
8. What are the potential complications for this patient?
9. Four months after laminectomy and bracing, the patient presents with continued thoracic pain, what is your initial diagnostic work-up?
10. What would be your operative approach for this scenario?

## Answers

1. *What is your differential diagnosis?*
   - Osteomyelitis/discitis
     - Pyogenic
     - Tuberculosis
     - Fungal
     - Epidural abscess
   - Metastatic bone cancer
     - Lung
     - Prostate
     - Kidney
     - Thyroid
   - Multiple myeloma with associated plasmacytoma
   - Thoracic disc herniation associated with Modic changes
   - Spinal meningioma
   - Primary bone cancer
     - Osteochondroma
     - Osteoid osteoma
     - Chondrosarcoma
     - Chordoma
2. *What initial imaging or lab tests would you order?*
   - The initial laboratory and imaging investigation should include[1,2]:
     - Complete blood count with differential
     - Complete metabolic panel
       - Serum calcium
       - Blood urea nitrogen (BUN)/Cr
       - Liver function tests
     - Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)
     - Prothrombin time/international normalized ratio/activated partial thromboplastin time (PT/INR/aPTT)
     - Blood cultures
     - Plain films of spine
     - MRI with and without contrast of the cervical and thoracic spine
     - CT scan of the cervical and thoracic spine
   - Laboratory examination revealed WBC = 19.8, mature granulocytes = 91%, Hgb = 8.9, PLT = 745, BUN = 31, Cr = 1.62 (1.64 on previous visit), calcium = 8.6, CRP = 20.5, other electrolytes were normal, and LFTs were slightly elevated. One of the two blood cultures and wound culture grew 1+ methicillin-resistant *Staphylococcus aureus* (MRSA).
3. *Interpret the MRI findings.*
   - T1-weighted sagittal MRI (▶**Fig. 135.1**b) of the thoracic spine demonstrates hypointensity within the T2 and T3 vertebral bodies. There is an isointense ventral epidural mass extending from T1 to T3 causing severe spinal cord compression.
   - Contrast-enhanced images (▶**Fig. 135.1**a) demonstrate an avidly enhancing lesion at T2–T3 level.
   - The T2-weighted sagittal MRI (not shown here) demonstrates hyperintensity of the T2–T3 disc resulting in displacement of the spinal cord and severe spinal stenosis.
4. *What are the radiographic elements of osteomyelitis/discitis on MRI?*
   - One major radiographic element in osteomyelitis/discitis is bone marrow edema, though it is not present in all patients. Bone marrow edema may be more easily apparent on a short-tau inversion recovery (STIR) or fat-suppressed T2-weighted sequences.
   - Standard T1 with and without contrast can detect inflammatory changes as well.
   - Other findings suggestive of osteomyelitis/discitis are paradiscal inflammation, vertebral erosions, intradiscal fluid, unusual endplate and vertebral enhancement, and/or abscess formation.
   - Modic degeneration (type 1) has been reported in some patients with osteomyelitis, but its presence on imaging may suggest another diagnosis. Liquid purulent abscess can be distinguished from phlegmon by the former having ring enhancement with surrounding hypointense core as opposed to phlegmon which usually appears to have uniform enhancement.
5. *What are the risk factors for this condition?*
   - The risk factors for pyogenic or any other form osteomyelitis/discitis are[3,4]:
     - Age > 50 years
     - Intravenous drug use
     - Prior spinal surgery
     - Prior steroid injection/interventional pain procedures
     - Endocarditis
     - Rheumatoid arthritis
     - End stage renal disease
     - Diabetes
     - Vasculopathy
     - Gender: males
     - Immunocompromised states (human immunodeficiency virus [HIV] infection, transplant patients)
     - Alcohol abuse
6. *What are the common modes of transmission for osteomyelitis/discitis?*
   - Mechanisms of transmission include[5,6]:
     - Hematogenous spread from bacteremia through Batson's plexus
     - Contiguous spread or direct inoculation—associated with pain procedures, spinal instrumentation, or prior surgery
7. *Describe indications for surgical interventions that may be necessary in a patient who has osteomyelitis/discitis.*
   - There are six principles that guide surgical intervention in osteomyelitis/discitis[1]:
     - Neurological deficits

## Answers (*continued*)

  - Intractable pain
  - Sepsis
  - Surgical biopsy
  - Medical treatment failure
  - Spinal deformity or instability

  This patient was initially managed with decompressive laminectomy due to myelopathy as well as brace and antibiotics consisting of 6 weeks of vancomycin.

8. *What are the potential complications for this patient?*
   - Before antibiotics, mortality rate was 25% compared to current morality rate of less than 11%.[3]
   - One study found that the relapse rate for osteomyelitis was about 12% determined by clinic and laboratory date after medical or surgical intervention, with most occurring within a few months after treatment.[7] The risk of relapse can be assessed by the presence or absence of certain risk factors such as bacteremia, age, associated abscesses, delayed diagnosis, or specific spinal lesion location.[7]
   - Postsurgical relapse was 17 to 35% and depended on the surgical approach selected.[8] The lowest postsurgical relapse was associated with the anterior fusion-posterior fixation with implants.[8]
   - This patient is exhibiting some of the featured complications of osteomyelitis such as neurological deficits and abscess formation.
9. *Four months after laminectomy and bracing, the patient presents with continued thoracic pain, what is your initial diagnostic work-up?*
   - ESR and CRP to determine if normalization occurred, and hence control of the inflammatory response to the infection
   - 36″ standing scoliosis films
   - May also need to obtain CT and MRI of the thoracic spine to assess the status of the bony and neurological elements at that time.
10. *What would be your operative approach for this scenario?*
    - It is the authors' opinion that a posterior vertebral column resection with potential removal of the T3 vertebral body (or pedicle subtraction osteotomy) and long segment posterior instrumented fusion would be an ideal option to treat this condition due to the ability to decompress as well as obtain anterior column restoration through a familiar approach (see ▸**Fig. 135.2** for a postoperative X-ray).
    - This may not be the only approach available to treat this condition; however, several arguments are in disfavor of alternate approaches: An anterior approach would require division of manubrium and mobilizing great vessels. Furthermore, placement of anterior plate would require an en face view which would be difficult to obtain in an already kyphotic spine. Additional risk of chylous leak exists. A posterolateral thoracotomy would require mobilizing and elevating the scapula, and require leaving a chest tube in most cases.

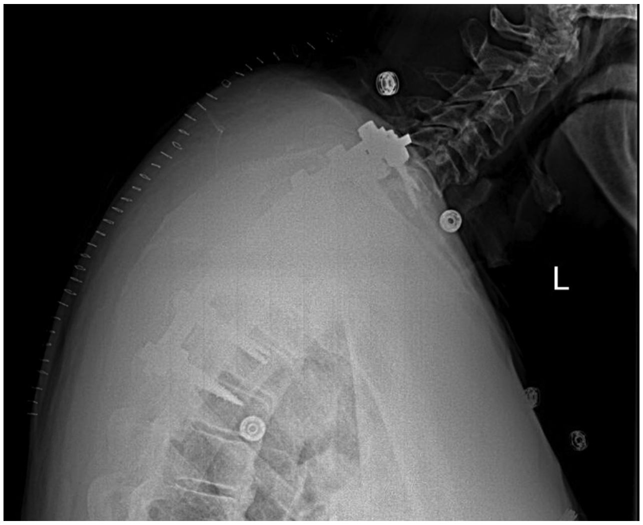

**Fig. 135.2** Lateral radiograph obtained postoperatively.

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# Case 136 Spinal Cord Inflammatory Disorder

*Vishal Patel, Rahul Shah, Achal Patel, and Juan Ortega-Barnett*

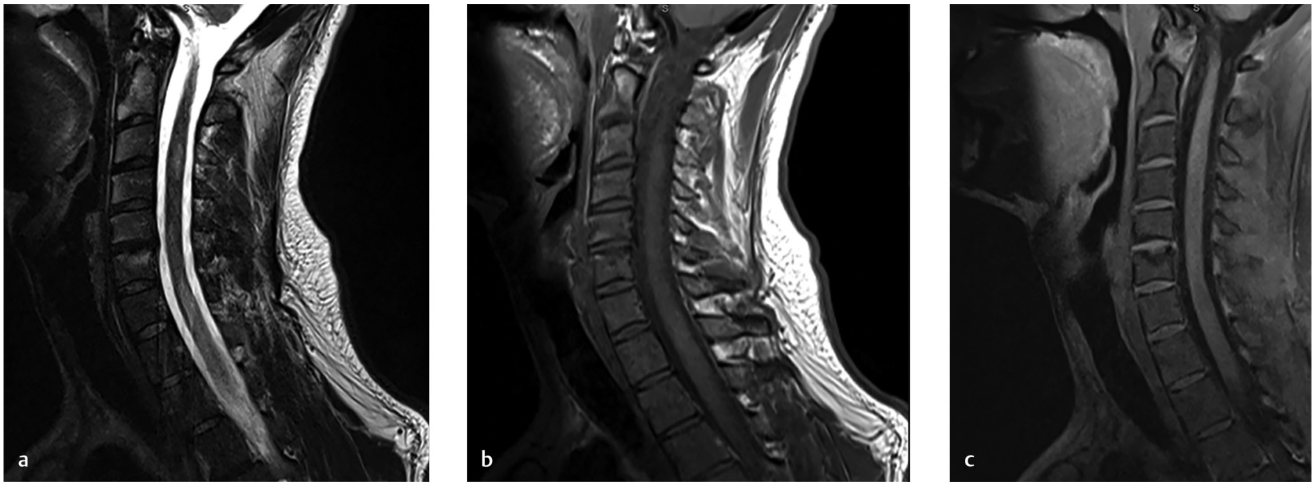

**Fig. 136.1** MRI of the cervical spine prior to treatment. (**a**) T2-weighted, (**b**) T1-weighted without contrast, and (**c**) T1-weighted with contrast.

## Clinical Presentation

- A previously healthy 31-year-old male presents with progressive bilateral lower extremity weakness and urinary incontinence. He states his symptoms began 1 week prior and was associated with fever, sweating, generalized lethargy, and nonradiating lower back pain.
- He mentions that he started having difficulty urinating 2 days ago. Today, he is not able to void or walk. He denies vision changes, trauma, recent infections, or recent travel.
- Neurologic examination is significant for 1/5 strength and decreased sensation to light touch, pain, and temperature in the lower extremities bilaterally. Deep tendon reflexes are 2+ throughout. The rest of his physical exam is normal.

## Questions

1. What is your differential diagnosis?
2. How will you work up this patient?
3. Cerebrospinal fluid (CSF) work-up is negative except for elevated IgG index with pleocytosis. Metabolic and serology studies are normal. Interpret these CSF findings and the MRI in ▶**Fig. 136.1**.
4. How does transverse myelitis (TM) present clinically?
5. List causes of secondary (disease-associated) TM.
6. Briefly outline the diagnostic criteria for idiopathic acute TM.
7. Differentiate TM from Guillain–Barré syndrome (GBS).
8. What is the most common level of sensory deficit?
9. What treatment options can you offer to a patient with TM?
10. What is the prognosis for patients with TM following treatment?

## Answers

1. *What is your differential diagnosis?*
   - Compressive (intervertebral disc herniation, hematoma, neoplasm)
   - Vascular (anterior spinal artery infarction, spinal-dural arteriovenous fistula)
   - Metabolic (vitamin B12 deficiency, copper deficiency, chronic liver disease)
   - Inflammatory/autoimmune (TM, multiple sclerosis, GBS)
2. *How will you work up this patient?*
   - Basic blood work (complete blood count [CBC], chemistry panel, renal function, coagulation studies, liver function tests)
   - Metabolic labs (vitamin B12 and copper levels)
   - Serology studies (enzyme-linked immunosorbent assay (ELISA), antinuclear antibody (ANA), antiphospholipid antibodies, rheumatoid factor, anti-dsDNA antibodies)
   - Gadolinium-enhanced and noncontrasted MRI of the entire spine
   - CT myelography is a reasonable alternative to rule out compressive lesions if MRI is contraindicated.
   - Lumbar puncture for CSF studies: in the absence of contraindications on imaging studies (cell count and differential, Gram stain, protein, glucose, aerobic/anaerobic/fungal cultures and sensitivities, acid-fast stain, oligoclonal bands, IgG index, viral studies, and cytology). Recall that up to one-third of CSF studies will be normal in the acute phase of the condition.
   - MRI brain to identify potential subclinical inflammatory or demyelinating processes.[1,2]
3. *Cerebrospinal fluid (CSF) work-up is negative except for elevated IgG index with pleocytosis. Metabolic and serology studies are normal. Interpret these CSF findings and the MRI in* ▶**Fig. 136.1**.
   - MRI shows T2 signal hyperintensity within the spinal cord and enhancement on T1 with gadolinium. No compressive lesion or vascular malformation is seen.
   - Given these imaging findings, CSF studies, and clinical presentation, TM is the likely diagnosis and a thorough work-up should be completed to find any secondary causes responsible, to differentiate between idiopathic and disease-associated TM.
   - In the absence of such CSF and MRI findings, consider noninflammatory causes such as anterior spinal artery infarction, arteriovenous fistula, radiation myelitis, or fibrocartilaginous embolism.[3]
4. *How does transverse myelitis (TM) present clinically?*
   - Common symptoms include back pain, limb weakness, sensory changes, and autonomic dysfunction (impaired bowel and bladder function, paroxysmal hypertensive episodes).[4,5]
   - Patients often report acute onset of symptoms that worsen over several days to weeks.
   - Symptoms are typically bilateral and a clearly defined sensory level is often present.[2]
5. *List causes of secondary (disease-associated) TM.*
   - Infection: herpes simplex virus (HSV), cytomegalovirus (CMV), Epstein–Barr virus (EBV), tuberculosis, Lyme disease, syphilis, West Nile virus
   - Autoimmune: systemic lupus erythematosus (SLE), mixed connective tissue disease, Sjögren's syndrome, scleroderma
   - Demyelinating: multiple sclerosis, neuromyelitis optica (NMO), acute disseminated encephalomyelitis (ADEM)[1]
6. *Briefly outline the diagnostic criteria for idiopathic acute TM.*
   - Inclusion criteria[2]
     - Development of sensory, motor, or autonomic dysfunction attributable to the spinal cord
     - Bilateral signs/symptoms with defined sensory level
     - Exclusion of compressive etiologies
     - Inflammation within the spinal cord demonstrated by CSF pleocytosis, elevated IgG index, or gadolinium enhancement on MRI
     - Progression to nadir between 4 hours to 21 days following onset of symptoms
   - Exclusion criteria
     - Previous radiation to the spine
     - Clear clinical deficit attributable to anterior spinal artery thrombosis
     - Presence of arteriovenous malformations
     - Serologic or clinical evidence of connective tissue disorders (e.g., sarcoidosis, Behcet's disease, SLE)
     - Central nervous system (CNS) manifestations of bacterial or viral disease
     - Brain MRI abnormalities suggestive of multiple sclerosis
     - History of clinically apparent optic neuritis
7. *Differentiate TM from Guillain–Barré syndrome (GBS).*
   - TM is often confused with GBS.
   - Paraparesis or quadriparesis (if cervical lesion), early loss of bowel/bladder control, hyperreflexia (although hyporeflexia may be seen in acute stages), gadolinium enhancement on MRI spine, CSF pleocytosis, and increased CSF IgG index supports the diagnosis of TM.
   - Ascending weakness and sensory changes in a glove and stocking distribution, hyporeflexia, cardiovascular autonomic dysfunction, normal MRI spine, and elevated CSF protein in the absence of pleocytosis (albuminocytologic dissociation) favor the diagnosis of GBS.[6]

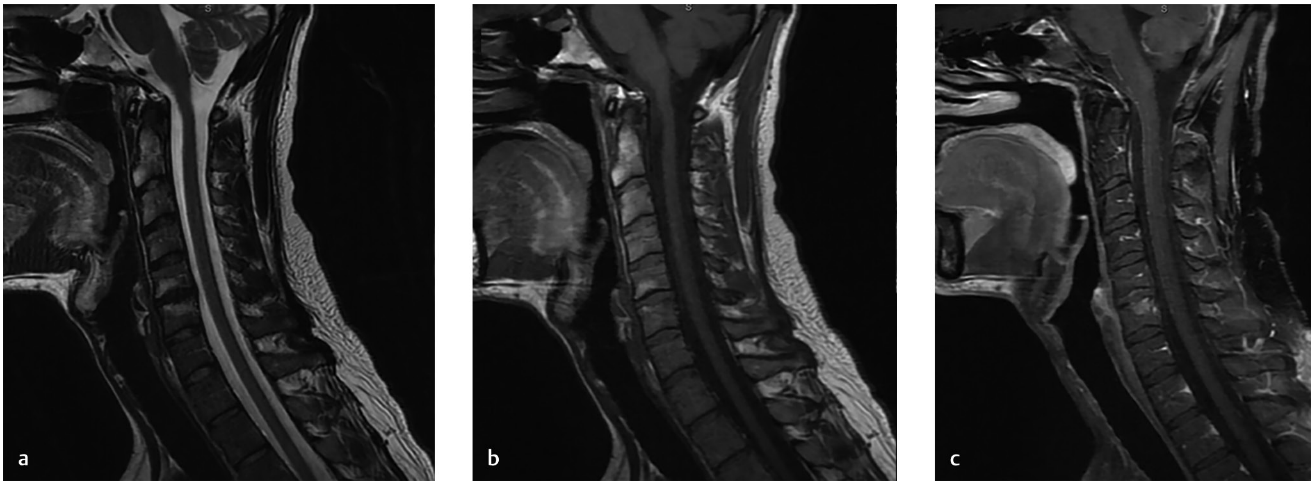

**Fig. 136.2** MRI of the cervical spine one month after treatment. (**a**) T2-weighted, (**b**) T1-weighted without contrast, and (**c**) T1-weighted with contrast.

## Answers (*continued*)

8. *What is the most common level of sensory deficit?*
    - Approximately 80% of patients present with a thoracic sensory level.[7]
9. *What treatment options can you offer to a patient with TM?*
    - High-dose methylprednisolone 1000 mg IV should be given for 3 to 7 days in the setting of an inflammatory/demyelinating etiology.
    - Plasma exchange can be considered if there is suboptimal response to steroids.[8]
    - Repeat MRI should be done after treatment.
    - The neurology service can be consulted for additional management strategies.
    - Admission to the hospital with Foley catheter placement, deep venous thrombosis (DVT) prophylaxis, and physical and occupational therapy consultation to avoid contractures.
10. *What is the prognosis for patients with TM following treatment?*
    - One-third of patients recover with no residual deficits, one-third retain moderate disabilities, and one-third show no improvement.
    - Recurrence of symptoms is more common among patients with secondary TM.[7,9]

    In the present case, the patient recovered with no significant residual deficits. Posttreatment MRI is shown in ▶**Fig. 136.2**.

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# Case 137 Chiari I Malformation

*Mahmoud AlYamany, Homoud AlDahash, and Abdulrahman J. Sabbagh*

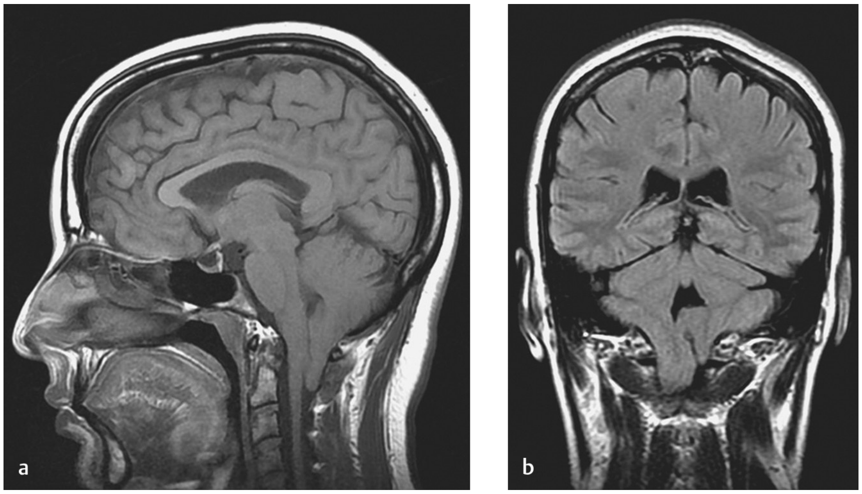

**Fig. 137.1** (**a**) Sagittal and (**b**) coronal T2-weighted MR images, demonstrating small posterior fossa, descended cerebellar tonsils down to the level of C1–C2 level posteriorly, and compression of the craniocervical junction and cervical spinal cord.

## Clinical Presentation

- A 32-year-old woman presents with a long history of neck pain, intermittent bilateral shoulder pain worsened with neck flexion and extension and during coughing and/or sneezing.
- She also complains of intermittent numbness in the tips of her fingers bilaterally.
- On physical examination, she has no neurologic deficits.
- Her past medical history is remarkable for an operation as a child for scoliosis, which was corrected at the age of 13 years. She did not have any further problem in that regard.
- MRI is obtained and shown in ▶**Fig. 137.1**.

## Questions

1. What is the differential diagnosis?
2. What does the sagittal image of the MRI demonstrate?
3. Describe your management.
4. What is the expected outcome?
5. What are the possible complications of this procedure?

   Assume this patient presented with progressive difficulty walking and spasticity. If she also presented with a spine MRI showing a syrinx in the upper thoracic and lower cervical cord, what would be your answers to the following questions?
6. What is the best management for the syrinx?
7. What are the pathogenesis theories of syringomyelia in Chiari I malformation?
8. Would you expect radiologic or clinical improvement to occur first?
9. How long would you wait before you expect the syrinx to start narrowing?
10. What are the different types of Chiari malformations?

## Answers

1. *What is the differential diagnosis?*
   - Differential diagnosis includes:
     - Hydrocephalus
     - Chiari malformation with cervical spinal cord syrinx
     - Isolated cervical spinal cord syrinx following scoliosis surgery
     - Cervical spine spondylosis
     - Tumors of the cervicomedullary junction or cervical spine
2. *What does the sagittal image of the MRI demonstrate?*
   - Sagittal MRI of the brain demonstrates:
     - Small posterior fossa
     - Descended cerebellar tonsils, below the level of the craniocervical junction
     - Secondary myelomalacia of the cervical spinal cord
3. *Describe your management.*
   - Management consists of suboccipital craniotomy.
     - Removing the rim of the foramen magnum and the posterior arch of C1
     - This is followed by duraplasty, with or without arachnoid lyses and/or tonsillar resection.
4. *What is the expected outcome?*
   - The expected outcome would be a stabilization of symptoms or arrested symptom progression.
   - There is a chance of improvement in her neurologic condition and a good chance of improvement of the headache and neck and shoulder pain.[1–3]
5. *What are the possible complications of this procedure?*
   - Possible complications of the procedure include the following[4]:
     - Cerebrospinal fluid (CSF) leak
     - Subdural hygroma
     - Wound infection
     - Further herniation of the tonsils due to raised intracranial pressure
     - There is a small chance of intraoperative neural tissue injury.
     - Postoperative headache: causes include chemical meningitis from the patch or glue
     - Possibility of nocturnal respiratory depression following posterior fossa manipulation (more common in Chiari type II)
6. *What is the best management for the syrinx?*
   - First line of management is treating the Chiari malformation via a suboccipital decompression[1] (see answer of Question 3 for details).
   - Alternative options include adding a syringosubarachnoid shunt after the Chiari decompression in selected cases (presence of a large syrinx, with significant thinning of the spinal cord tissue and obliteration of the spinal subarachnoid space, especially when combined with syrinx-related symptoms),[5] or syringopleural shunt.
7. *What are the pathogenesis theories of syringomyelia in Chiari I malformation?*
   - Several theories exist:
     - Gardner's hydrodynamic theory[6]:
       - ○ First to recognize the frequent association of syringomyelia with Chiari I malformation
       - ○ Delay in the perforation of the roof of the rhombencephalon
       - ○ Subarachnoid space does not fully open.
       - ○ CSF becomes trapped in ventricular system.
       - ○ The exaggerated pulsations are directed into the central canal causing hydromyelia.
     - Williams theory[7]:
       - ○ Postulated and later confirmed that such an obstruction could act as a valve, allowing CSF to cross the foramen magnum rostrally more effectively than caudally.
       - ○ Craniospinal dissociation is described where CSF may be "sucked" from the fourth ventricle into the central canal.
     - Ball and Dayan theory[8]:
       - ○ Activities that increase thoracic or abdominal pressure such as coughing and straining cause the spinal CSF to be diverted into the spinal cord parenchyma along dilated Virchow–Robin spaces.
     - Aboulker theory[9]:
       - ○ Spinal CSF enters into the cord by way of the dorsal root entry zone.
       - ○ Absorption occurs either by
         - ♦ Blood vessels of the spinal gray matter
         - ♦ Rostral drainage through the central canal into the fourth ventricle
     - Oldfield theory[10,11]:
       - ○ The force that pushes spinal subarachnoid CSF into the cord parenchyma is the CSF pulsation pressure occurring during the cardiac cycle.
       - ○ This pulsation will drive CSF into the cord along perivascular spaces.
     - Other theories such as Greitz[12] (intramedullary pulse pressure theory):
       - ○ Suggests that syringomyelia is caused by increased pulse pressure in the spinal cord and that the syrinx consists of extracellular fluid rather than CSF
8. *Would you expect radiologic or clinical improvement to occur first?*
   - Expect to see improvement in clinical symptoms before radiologic improvement.
9. *How long would you wait before you expect the syrinx to start narrowing?*
   - The radiologic improvement in the width of the syrinx is expected within the first 3 to 6 months, but continued narrowing of the syrinx may take years.

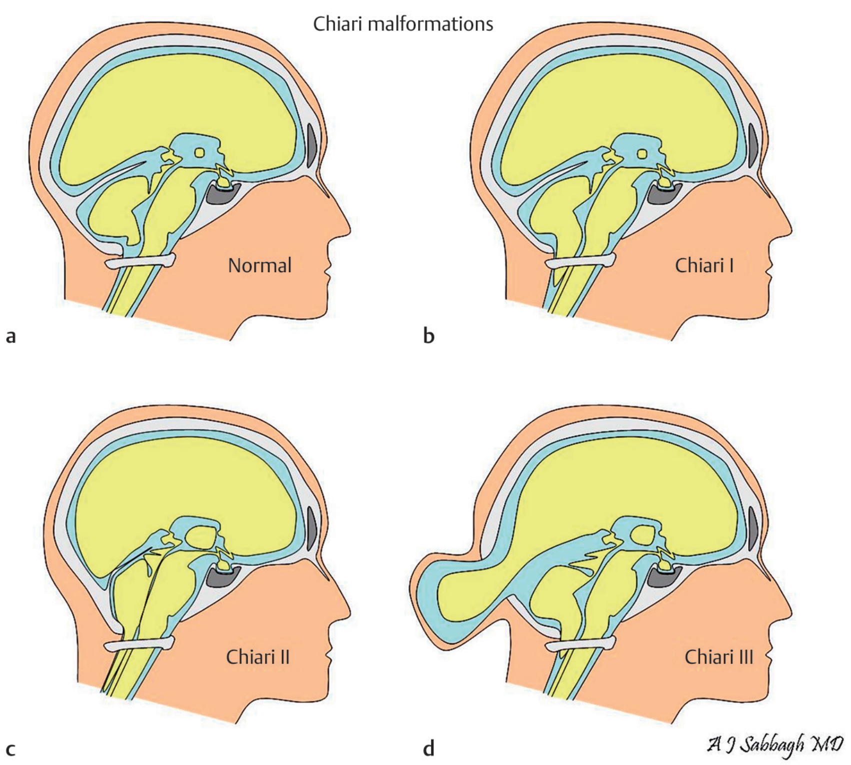

**Fig. 137.2** Diagrams demonstrating normal posterior fossa appearance and variants of Chiari malformations on sagittal planes. (**a**) Normal posterior fossa anatomy. (**b**) Chiari malformation type I: shallow posterior fossa, tonsillar descent down to the level of C1–C2 junction, and posterior compression of the cervical spinal cord. (**c**) Chiari malformation type II: shallow posterior fossa, descent of the cerebellar hemispheres, vermis, and tonsils to the upper cervical spinal canal level, and posterior compression of the brainstem and the cervical spinal cord. (**d**) Chiari malformation type III: shallow posterior fossa, tonsillar herniation, brainstem and cervical spinal cord compression, and associate occipital encephalocele.

## Answers (*continued*)

- According to Wetjen et al's series of 29 patients with syringomyelia and Chiari I malformations, the average diameter of the syrinx preoperatively was ~6.9 mm (with a 2.1 mm standard deviation), whereas postoperatively it was less than 1.5 mm at last evaluation ($p < 0.0001$).
- The median time for the syrinx narrowing is 3.6 months after a Chiari decompression is completed.[11]

10. *What are the different types of Chiari malformations?*
    - There are four types of Chiari malformations.[13]
    - See ▶**Fig. 137.2** for details of each of the three main types.
    - Note that Chiari type IV includes cerebellar hypoplasia.[13]

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# Section VII Peripheral Nerve Pathology

# Case 138 Median Nerve Entrapment at the Wrist

*Jonathan Yun and Christopher J. Winfree*

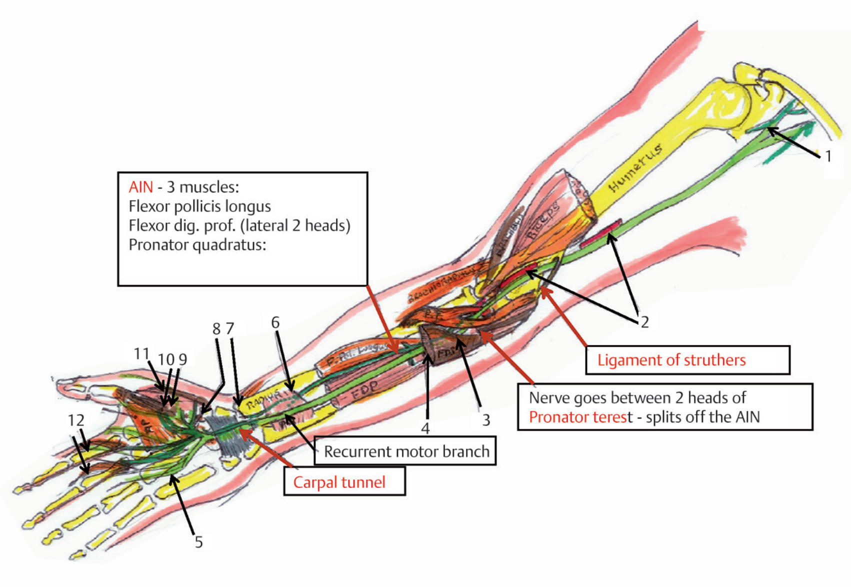
AIN - 3 muscles:
Flexor pollicis longus
Flexor dig. prof. (lateral 2 heads)
Pronator quadratus:
Humerus
Biceps
Brachialis
Brachioradialis
P.T.
FDS
F. Pol. Longus
FDP
Radius
AP
1
2
3
4
5
6
8 7
11
10 9
12
Ligament of struthers
Nerve goes between 2 heads of
Pronator terest - splits off the AIN
Recurrent motor branch
Carpal tunnel

**Fig. 138.1** Artist's rendering of median nerve anatomical course and locations of entrapment. Sites of entrapment are highlighted in *pink*. AIN, anterior interosseous nerve; FDP or Flexor Dig. Prof., flexor digitorum profundus; PQ, pronator quadratus; AP, adductor pollicis; PT, pronator teres; F. Pol. Longus, flexor pollicis longus; FDS, flexor digitorum superficialis. (1) Lateral and medial cords join to form the median nerve. (2) Median nerve descends along the medial edge of the brachial artery. (3) Median nerve passes under the flexor digitorum superficialis (arcade forms the sublimis bridge). (4) Median nerve descends between flexor digitorum superficialis and profundus. (5) Sensory branches of the median nerve. (6) Anterior interosseous nerve (AIN) reaches deep to the pronator quadrates. (7) AIN gives off the wrist articular branches. (8) Recurrent motor branch variation. (9) Abductor pollicis. (10) Flexor pollicis. (11) Opponens pollicis. (12) Lumbricals I and II.

## Clinical Presentation

- A 55-year-old right-handed woman with a history of diabetes mellitus and hypothyroidism, presents with a 4-month history of "pins and needles" in her first three fingers, notably sparing the fifth digit, in her dominant hand. She reports some subjective weakness of her right thumb.
- Episodes are transient, mildly painful, and typically occur at night awakening her from sleep. Some relief is found after awakening and shaking the hand for a few minutes.

## Questions

1. What is the differential diagnosis?
2. What are possible locations of median nerve entrapment?
3. What is the most likely diagnosis?
4. What special physical exam findings and maneuvers would support this diagnosis?
5. What diagnostic tests should be ordered?
6. What nonsurgical measures can be attempted to ameliorate the patient's symptoms?
7. When should surgery be considered?
8. What are the surgical options?
9. What complications are associated with carpal tunnel release?

## Answers

1. *What is the differential diagnosis?*
   - Cervical radiculopathy (C6–C7)
   - Carpal tunnel syndrome (CTS)
   - Brachial plexopathy
   - Supracondylar process syndrome (Struthers' ligament)
   - Pronator teres syndrome
   - Anterior interosseous nerve (AIN) syndrome
2. *What are possible locations of median nerve entrapment?*
   - ▶ **Fig. 138.1** shows the anatomic localizations of median nerve compression sites.
3. *What is the most likely diagnosis?*
   - CTS results from median nerve compression at the carpal tunnel formed by the carpal bones and ligaments as the lateral walls and floor and the transverse carpal ligament as its roof.[1]

## Answers (*continued*)

- Risk factors include repetitive hand movements, pregnancy, diabetes mellitus, acromegaly, hypothyroidism, multiple myeloma, amyloidosis, mucopolysaccharidosis, and rheumatoid arthritis.
- Women are twice as likely to be affected as men, and symptoms often arise during middle age.
- Classic symptoms are restricted to a median nerve distribution distal to the carpal tunnel, but patients can present with proximal arm pain as well.

4. *What special physical exam findings and maneuvers would support this diagnosis?*
   - Phalen's test: patient rests elbows on exam table with forearms upright and allows wrist flexion with gravity assistance. Considered positive if median nerve distribution paresthesias commence or increase within 60 seconds.
   - Tinel's sign: percussion of the median nerve at the wrist. Considered positive if reproduces paresthesias in a median nerve distribution.
   - Durkan's compression test: application of continuous pressure to the median nerve at the transverse carpal ligament produces median nerve paresthesias, which are relieved by release of pressure.
   - Sensory: for example, two-point discrimination, monofilament testing, vibration detection, sensory abnormalities should spare the palmar cutaneous distribution, since that nerve arises from the median nerve proximal to the carpal tunnel.
5. *What diagnostic tests should be ordered?*
   - Electromyography and nerve conduction studies (EMG/NCS) are the standard diagnostic tests for CTS; conduction slowing of the median nerve across the wrist and changes consistent with denervation in the distal median-innervated muscles are commonly seen in CTS.
   - Cervical spinal imaging studies are unnecessary to make the diagnosis of CTS but are helpful to exclude other problems in the differential diagnosis such as cervical radiculopathy.
   - In this case, physical exam and NCS were consistent with the diagnosis of right-sided CTS; MRI of the cervical spine was unremarkable.
6. *What nonsurgical measures can be attempted to ameliorate the patient's symptoms?*
   - Nonsurgical measures should be attempted first in an effort to avoid surgery.
   - Behavioral and environmental modifications, including ergonomics associated with precipitating activities and avoidance of repetitive, exacerbating movements.[2]
   - Wrist splinting, which keeps the hand in a neutral or slightly extended position, can be helpful, especially at night when patients have a tendency to sleep with the wrists flexed, thus aggravating their symptoms.
   - Local injection of corticosteroids into carpal tunnel can be an effective temporizing measure, especially useful in patients trying to avoid surgery in the setting of pregnancy. Antiepileptics such as gabapentin and pregabalin may also be tried.
   - Nonsteroidal anti-inflammatory medications, diuretics, vitamin supplements, and chiropractic manipulation are not thought to be helpful.[3]
7. *When should surgery be considered?*
   - Persistence of symptoms that do not improve with conservative management, typically longer than 1 month.[3]
   - Prolonged symptoms, presence of a positive Tinel's sign, and evidence of thenar wasting are associated with poorer outcomes with conservative management.[4]
   - Early evidence of conduction slowing along the median nerve at the wrist on NCS.
   - Weakness of median-innervated muscles support need for surgical decompression.
   - Denervational changes on needle EMG of median-innervated muscles distal to the carpal tunnel need not be present, but prompt earlier surgical intervention if present.
8. *What are the surgical options?*
   - Overall, surgical treatment of CTS results in significantly improved symptoms over conservative management.[5]
   - Mini-open carpal tunnel release involves decompressing the median nerve at the wrist through a 2-cm incision; although the patients may gently use the operated hand almost immediately, the incision requires a month or so of healing prior to aggressive work or rehabilitation activities (▶ **Fig. 138.2** and ▶ **Fig. 138.3**).[6]
   - Endoscopic carpal tunnel release involves decompressing the median nerve at the wrist through two 5-mm incisions; recovery times prior to return to work or aggressive physiotherapy are shorter after the endoscopic release compared with open release.[6]
   - Endoscopic and open carpal tunnel release are equivalent in symptom relief and improving functional status and occurrence of major complications, though endoscopic release is associated with shorter return-to-work time[7] (▶ **Table 138.1**).

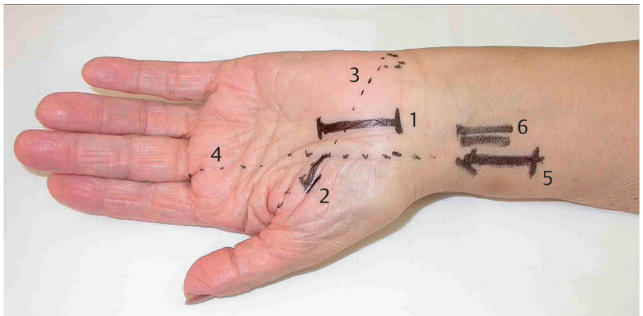

**Fig. 138.2** Intraoperative photograph of the patient featured in this case, illustrating the superficial landmarks, incision, and surgical anatomy of the mini-open carpal tunnel release. The incision (1) is roughly 2 cm in length and begins 1–2 mm distal to the wrist crease and in line with the radial aspect of the fourth digit. The recurrent motor branch of the median nerve (2) is located at the intersection of Kaplan's line, (3) drawn from the pisiform bone to the angle of the thumb, and the line drawn in line with the radial aspect of the third digit. (4) The palmar cutaneous branch of the median nerve (5) originates 5 cm proximal to the wrist crease along the radial aspect of the median nerve and generally traverses the wrist within its own fascial tunnel. The palmaris longus tendon (6) is sometimes seen proximal and just radial to the incision.

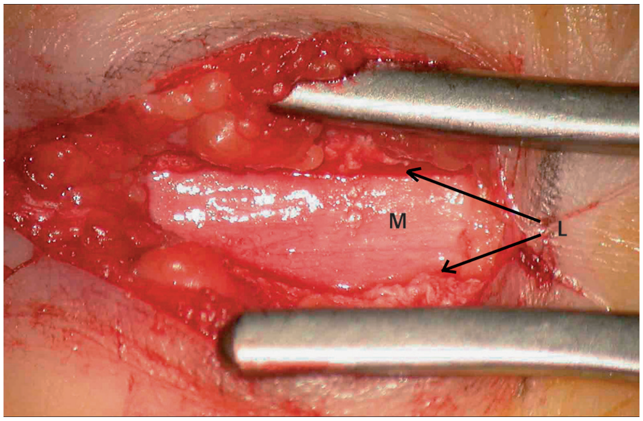

**Fig. 138.3** Intraoperative photograph of the patient featured in this case, illustrating the median nerve (*M*) following division of the transverse carpal ligament (*L*). Elevation of the skin at each end of the incision by a small handheld retractor permits decompression of a total length of ~10 cm of median nerve through the 2 cm incision.

**Table 138.1** Endoscopic versus open carpal tunnel release meta-analysis summary[6]

| | Endoscopic | Open |
|---|---|---|
| **Symptom relief** | Similar to open | Similar to endoscopic |
| **Complications** | More transient neuropraxias | More wound infections, scarring, and tenderness |
| **Return to work** | 6 days earlier than open | 6 days later than endoscopic |

## Answers (*continued*)

9. *What complications are associated with carpal tunnel release?*
   - Injury to the main trunk median nerve, the palmar cutaneous branch, the thenar branch, common digital nerves, or contents of Guyon's canal[8,9]
   - Incomplete nerve decompression is the most common cause of treatment failure.
   - Partial or complete tenotomy of flexor tendons, resulting in rupture, adhesions, or triggering[9]
   - Injury to the superficial palmar arch or ulnar artery
   - Infection
   - Scar with tenderness
   - Postoperative pain syndromes including pain of the hypothenar/thenar eminence (pillar pain) or complex regional pain syndrome (CRPS)[9]

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# Case 139 Ulnar Nerve Compression at the Elbow

*Stephen M. Russell*

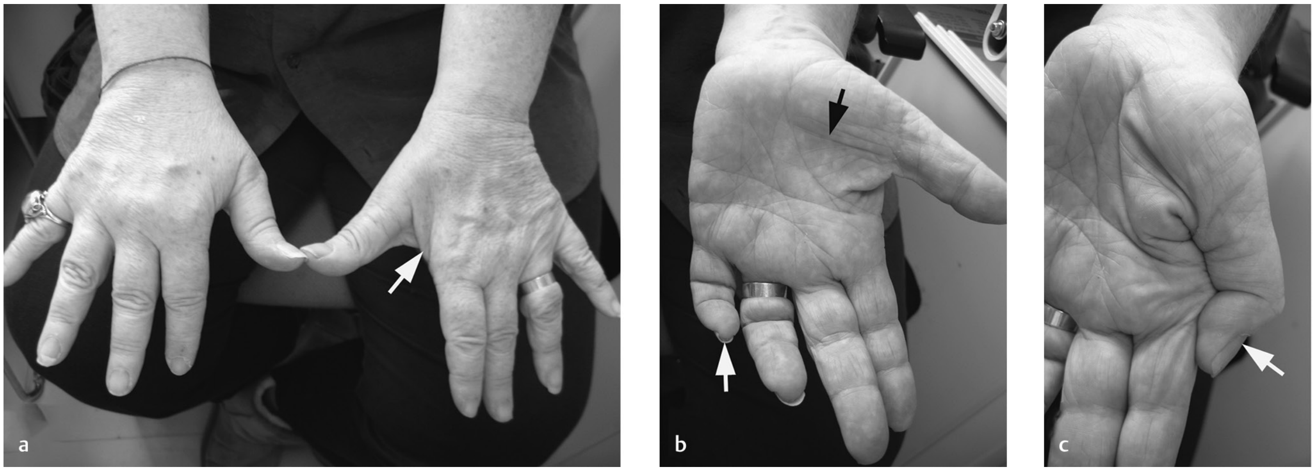

**Fig. 139.1** A patient with severe left ulnar nerve entrapment at the elbow with clinical signs (**a–c**).

## Clinical Presentation

- A 50-year-old woman presents with 6 weeks of elbow pain localized to the left medial epicondyle.
- It began after a prolonged period of holding the telephone with the affected hand.
- A few days later, she woke one morning with complete numbness in the small finger.
- Then over the ensuing weeks, she developed progressive hand atrophy, weakness, and incoordination.
- Her examination reveals dense numbness to light touch and pinprick on the hypothenar eminence (including the dorsum of the hand in this area), as well as the small finger and adjacent half of the ring finger.
- Severe atrophy of most hand intrinsic muscles is noted.
- Tapping in the retrocondylar groove causes paresthesias to occur in the small finger. ▶**Fig. 139.1** illustrates the patient's hands.
- She has no neck pain and no Spurling's sign.

## Questions

1. What is your differential diagnosis?
2. In ▶**Fig. 139.1**a, what atrophic muscle is labeled with the white arrow?
3. In ▶**Fig. 139.1**b, what atrophic muscle is labeled with the black arrow?
4. In ▶**Fig. 139.1**b, the white arrow indicates an examination finding that is commonly encountered with her diagnosis; what is it?
5. In ▶**Fig. 139.1**c, the white arrow indicates another examination finding that is commonly encountered with her diagnosis; what is it?
6. Describe the McGowan classification of ulnar nerve entrapment.
7. What are the nonoperative treatment options available?
8. What are the indications for surgery?
9. What are her surgical options for ulnar nerve decompression and/or transposition?
10. Describe the efficacy and common complications of ulnar nerve decompression and/or transposition.

## Answers

1. *What is your differential diagnosis?*
   - Her history and examination are classic for an ulnar nerve entrapment at the elbow, including her reporting a period of prolonged flexion that may have instigated her somewhat rapid progression of symptoms, a clear sensory loss in an ulnar nerve distribution, and the evidence of ulnar-innervated hand intrinsic atrophy.[1]
   - Other less likely diagnoses would include a C8 radiculopathy, an ulnar nerve entrapment at the wrist, and rarely, neurogenic thoracic outlet syndrome.
2. *In* ▶ **Fig. 139.1**a, *what atrophic muscle is labeled with the white arrow?*
   - There is marked atrophy of the first dorsal interosseous muscle.
   - Atrophy of this muscle is usually most obvious with moderate to severe ulnar nerve injury/ entrapment.
3. *In* ▶ **Fig. 139.1**b, *what atrophic muscle is labeled with the black arrow?*
   - Although most muscles of the thenar eminence are innervated by the median nerve, the largest one, the adductor pollicis, is innervated by the ulnar nerve; therefore, patients with ulnar neuropathy can also have atrophy of their thenar eminence.
4. *In* ▶ **Fig. 139.1**b, *the white arrow indicates an examination finding that is commonly encountered with her diagnosis; what is it?*
   - When the patient opens her affected hand, an “ulnar claw hand” is revealed. This consists of ring and small finger hyperextension at the knuckles (metacarpophalangeal joints) with superimposed interphalangeal joint flexion secondary to a tenodesis effect.
5. *In* ▶ **Fig. 139.1**c, *the white arrow indicates another examination finding that is commonly encountered with her diagnosis; what is it?*
   - A positive Froment’s sign
   - In patients with severe ulnar neuropathy, the adductor pollicis cannot adduct a straight thumb.
   - Instead, the patient flexes the distal interphalangeal joint (median-innervated flexor pollicis longus) to oppose the thumb against the hand.
6. *Describe the McGowan classification of ulnar nerve entrapment.*
   - Dr. McGowan classified ulnar nerve entrapment at the elbow as follows[2]:
     - Grade 1: purely subjective symptoms with mild hypesthesia
     - Grade 2: sensory loss with slight wasting and weakness of the hand intrinsic muscles
     - Grade 3: severe sensorimotor deficits
7. *What are the nonoperative treatment options available?*
   - Very few nonoperative treatments are available.
   - Some include wearing an elbow pad or an elbow splint; however, compliance with these is poor.
   - Physical and occupational therapy may be effective in resolving mild cases.
8. *What are the indications for surgery?*
   - Surgery is usually recommended for McGowan grade 2 and 3 ulnar nerve entrapments.
   - As a general rule, if there are no periods during the day where the hand is normal, then nerve damage is likely occurring and surgery should be an option.
   - Some patients with McGowan grade 1 entrapments also request surgery when other conservative measures have failed.
9. *What are her surgical options for ulnar nerve decompression and/or transposition?*
   - There are many surgical options for decompressing the ulnar nerve at the elbow (▶ **Fig. 139.2**).
   - One should use a technique they have personal experience with because, in general, most techniques are equally efficacious.
   - Simple in situ decompression of the ulnar nerve has become more popular as of late because of a few recent randomized trials concluding that a simple decompression is as effective as a transposition.[3–5]
10. *Describe the efficacy and common complications of ulnar nerve decompression and/or transposition.*
    - The results of ulnar nerve decompression and/or transposition are that 60 to 80% of patients improve, while the rest remain in the same condition, or occasionally get worse over time.[3–5]
    - Complications include wound pain, elbow numbness, and neuroma formation from injury to the posterior division of the medial antebrachial cutaneous nerve (▶ **Fig. 139.2**a).

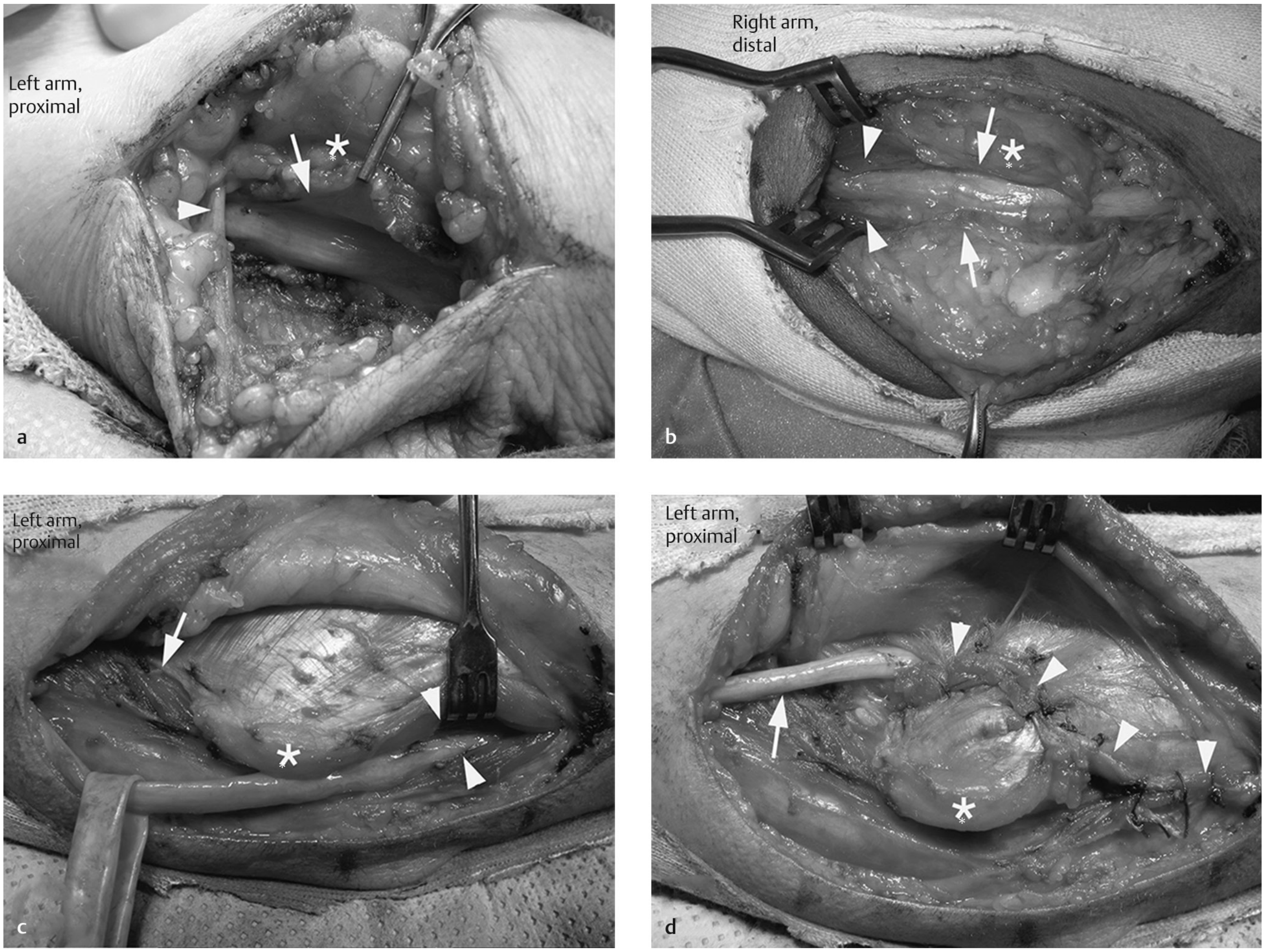

**Fig. 139.2** Intraoperative photographs of decompression and/or transposition techniques for ulnar neuropathy localized to the elbow. Arm laterality, orientation, as well as location of the medial epicondyle (*) are provided for each photograph. (**a**) A simple, in situ decompression of the ulnar nerve (same patient as ▶**Fig. 139.1**). Osborne's ligament has been transected and is held open by forceps (*white arrow)*. Although not visible, the ulnar nerve has been decompressed for 2 to 3 inches beyond both the proximal and distal exposures. The posterior branch of the medial antebrachial cutaneous nerve (*arrowhead)* often traverses the operative exposure and is at risk for iatrogenic injury. (**b**) Another simple decompression in a different patient. To release the cubital tunnel, both the two heads of the flexor carpi ulnaris (*arrowheads*) as well as the Osborne's ligament (*arrows*) have been released to decompress the ulnar nerve. (**c**) Exposure of the ulnar nerve prior to a submuscular transposition in a patient with moderate ulnar neuropathy. The ulnar nerve is fully neurolysed and is isolated with a Penrose drain. The heads of the flexor carpi ulnaris have been separated (*arrowheads*), and the medial intermuscular septum has been removed (*arrow*). Failure to remove this septum may lead to iatrogenic nerve compression when the ulnar nerve is subsequently transposed under the flexor–pronator mass. (**d**) Same patient as in (**c**). The ulnar nerve (*arrow*) has been placed under the flexor–pronator muscle mass, which has been sutured back together (*arrowheads*).

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# Case 140 Neurogenic Thoracic Outlet Syndrome

*Stephen M. Russell*

## Clinical Presentation

- A 32-year-old woman presents with 1-year history of left-hand weakness and incoordination, numbness in her medial left forearm, and paresthesia in her left fifth digit when she rotates her head to the right.
- Her weakness temporarily worsens with overhead arm activity, including combing her hair and reaching for items.
- She denies sensory symptoms in the thumb or first two fingers.
- She is a slim woman with a long neck and poor posture.
- On examination, she has mild atrophy and weakness in both the median- and ulnar-innervated hand intrinsic muscles compared with the opposite hand, hypesthesia along the medial left forearm, and a positive Tinel's sign with gentle tapping in the left supraclavicular space.
- Her radial pulse disappears in either arm when the arm is raised above her head.

## Questions

1. What is your differential diagnosis?
2. Describe the myotome and dermatome for C8, T1, and the lower trunk.
3. What diagnostic studies would you request to help confirm the diagnosis?
4. What are the different types of thoracic outlet syndromes?
5. What is the proposed pathophysiology of neurogenic thoracic outlet syndrome?
6. What are the treatment options?
7. What is the success rate of surgery for neurogenic thoracic outlet syndrome?

## Answers

1. *What is your differential diagnosis?*
   - Considering that her neurologic examination localizes her pathology to the lower trunk, the working diagnosis would be neurogenic thoracic outlet syndrome.
   - The differential diagnosis for neurogenic thoracic outlet includes cervical radiculopathy, carpal tunnel syndrome, ulnar nerve entrapment at the elbow, motor neuron disease, and a Pancoast tumor.[1]
2. *Describe the myotome and dermatome for C8, T1, and the lower trunk.*
   - Together, C8 and T1 innervate all of the hand intrinsic musculature via both the ulnar and median nerves (they also provide contribution to some more proximal muscles).
   - ▶**Fig. 140.1** demonstrates the combined dermatome of C8 and T1 which represents the region of possible numbness or paresthesias in patients with neurogenic thoracic outlet syndrome.
   - Classically, these patients have paresthesias and numbness along the medial forearm.
3. *What diagnostic studies would you request to help confirm the diagnosis?*
   - An apical lordotic radiograph of the cervical spine should reveal any cervical ribs or "beaked" C7 transverse processes.
   - MRI with and without contrast of the brachial plexus (which should include the cervical spine) is ordered to exclude a herniated disc, foraminal stenosis, and tumors.
   - Although frequently normal in mild cases, electrodiagnostic studies can reveal denervation in both ulnar- and median-innervated hand muscles (e.g., first dorsal interosseous and abductor pollicis brevis, respectively) and perhaps an absent nerve action potential from the medial antebrachial cutaneous nerve.
4. *What are the different types of thoracic outlet syndromes?*
   - Thoracic outlet is categorized as neurogenic, vascular, or disputed.[2]
   - Neurogenic and vascular thoracic outlet are both quite rare; the disputed type is much more common.
   - Neurogenic thoracic outlet syndrome requires clear demonstration of objective neurologic finding on examination or diagnostic tests, including atrophy, electrodiagnostic testing abnormalities, a cervical rib, and/or focal nerve swelling on MRI.
   - If no objective findings are present, the patient should be observed for their subsequent development.
5. *What is the proposed pathophysiology of neurogenic thoracic outlet syndrome?*
   - Most experts believe that accessory ligaments and/or fascial bands related to a cervical rib or "beaked"

## Answers (*continued*)

C7 transverse process compress and distort the brachial plexus in patients with neurogenic thoracic outlet syndrome.[3]

- It remains controversial whether the anterior scalene, per se, is responsible for nerve entrapment.

6. *What are the treatment options?*
   - For patients with mild symptoms and signs, posture training and a trial of physical therapy may lead to improvement.
   - If atrophy and/or weakness are present, then surgical decompression of the brachial plexus is indicated.
   - However, once profound and chronic atrophy is present, the chance of surgical decompression causing a substantial improvement is unlikely.
   - Therefore, early treatment is optimal.
   - An anterior suprascapular exposure of the brachial plexus is the preferred approach by neurosurgeons in patients with neurogenic thoracic outlet syndrome (▶**Fig. 140.2**).[4]
7. *What is the success rate of surgery for neurogenic thoracic outlet syndrome?*
   - If preoperatively the patient only has provocative symptoms (e.g., with overhead arm use), and only minimal signs of weakness and atrophy (i.e., mild cases), then surgical decompression leads to improvement in 85 to 90% of patients.
   - When significant atrophy and weakness are present (moderate-to-severe cases), the chance of partial improvement is approximately two-thirds.[5]

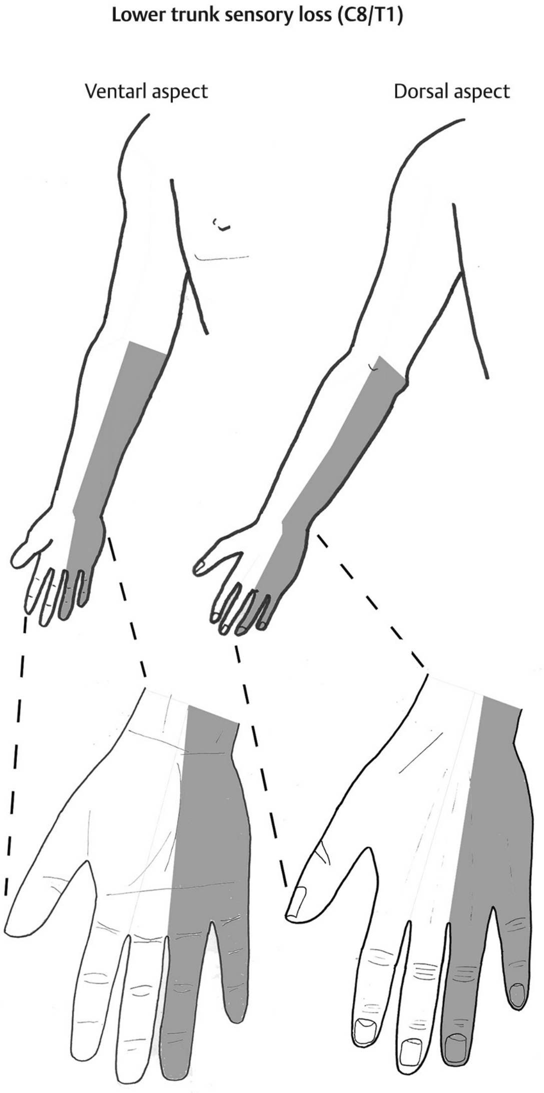

**Fig. 140.1** Location of lower trunk sensory loss (i.e., combined C8 and T1 sensory loss). Although variable, patients with neurogenic thoracic outlet syndrome often have sensory loss or paresthesias confined to the medial forearm (T1 dermatome), and less so in the small finger. (Reproduced from Russell 2007)[1]

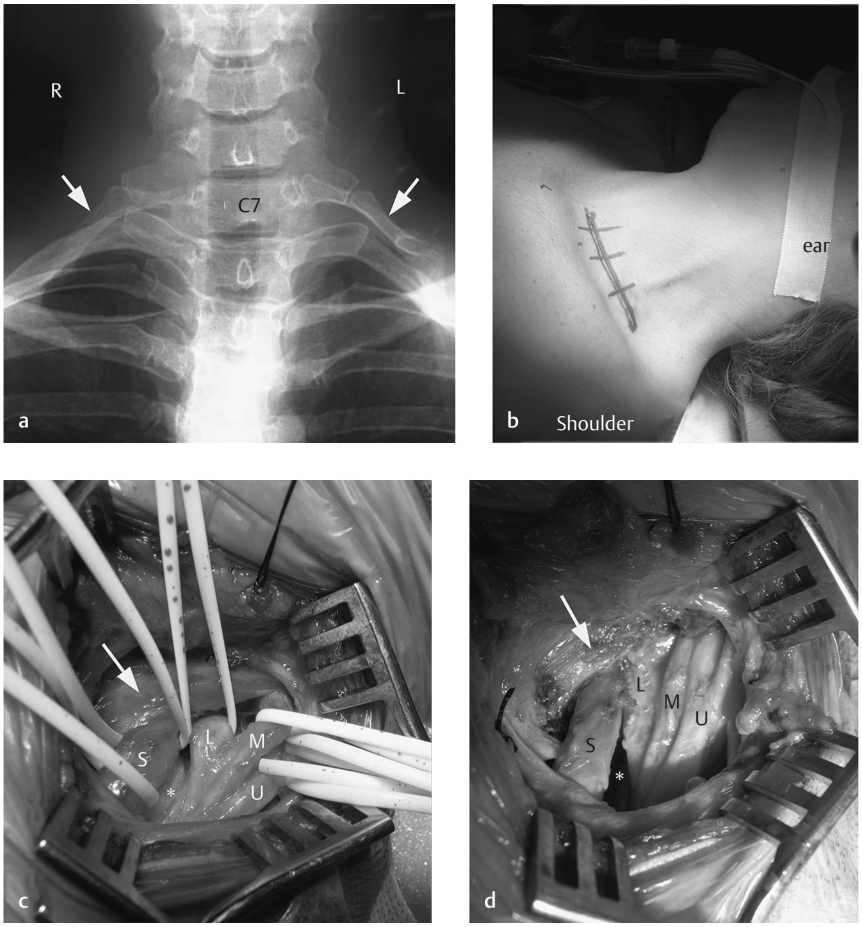

**Fig. 140.2** Woman with left-sided neurogenic thoracic outlet syndrome. (**a**) Apical lordotic radiograph demonstrating bilateral cervical ribs (*white arrows*). (**b**) The transverse incision used to expose the left-sided brachial plexus. Care must be taken to preserve the supraclavicular sensory nerves, which are subcutaneous and often traverse the operative field. (Subsequent operative images are in the same orientation as depicted here). (**c**) The brachial plexus (upper [U], middle [M], and lower [L] trunks), subclavian artery (S), cervical rib (*), and anterior scalene (*white arrow*) are all exposed via an anterior supraclavicular approach. The clavicle is to the left of the operative exposure; therefore, the subclavian artery and lower trunk are significantly stretched over the top of the cervical rib and displaced in a cranial direction. (**d**) The cervical rib and its underlying attachments to the first rib have been removed (*). A portion of the scalene muscle has also been removed. The neurovascular structures are now under no tension.

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# Case 141 Brachial Plexus Injury and Horner's Syndrome

*Hussam Abou-Al-Shaar, Perry S. Bradford, Christian A. Bowers, and Mark A. Mahan*

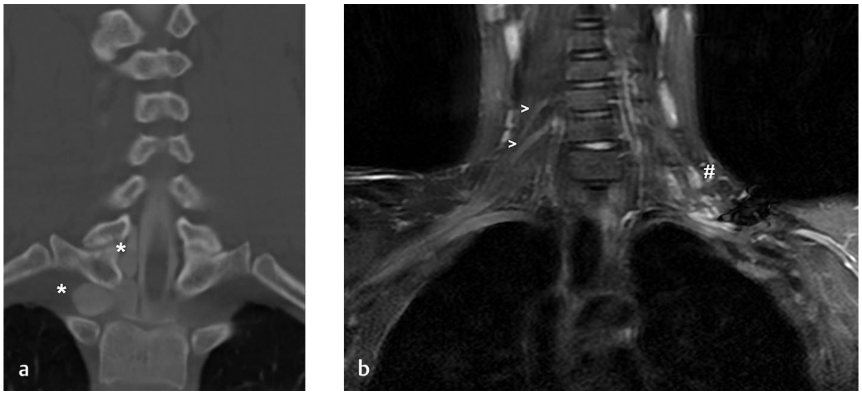

**Fig. 141.1** (**a**) CT myelogram of the cervical spine demonstrates pseudomeningoceles at C8 and T1 (*asterisks*). (**b**) Coronal T2-weighted MRI with fat suppression depicts hyperintensity of the upper, middle, and lower trunks on the left side (*pound sign*) consistent with rupture of the upper trunk, stretch injury to the middle trunk, and avulsion of the lower trunks (pseudomeningoceles not shown on this slice), in comparison to the normal fascicular intensity of the C5 and C6 roots on the patient's right side (*right brackets*).

## Clinical Presentation

- A 27-year-old right-handed man presents 4 months after a motorcycle collision with a flaccid left upper extremity. He has experienced persistent severe, refractory arm pain with episodic, "electric, burning, crushing" sensations in his otherwise insensate forearm and hand.
- Since the accident, he has had no spontaneous return of motor function in his arm and was able to only activate shoulder retraction and elevation on physical examination.
- Crude, ill-defined sensation was present on the lateral brachium and intact sensation in the armpit.
- Electromyography (EMG) demonstrated denervation injury with evidence of denervation and reinnervation (complex motor units) in the rhomboids. Nerve conduction testing found recordable sensory nerve action potentials in the ulnar nerve.
- Examination also revealed a positive Horner's syndrome: right eye (OD): – 4 mm, left eye (OS): – 2 mm.
- CT myelogram shows no obvious ventral nerve rootlets from C5 to T1, and MRI of the brachial plexus demonstrates extraforaminal discontinuity of C5, C6, and C7, with obvious avulsion of C8 and T1 (see ▸**Fig. 141.1**).

## Questions

1. What is the likely diagnosis?
2. What is the impact of the finding of anisocoria on your assessment?
3. What are the common injury patterns in supraclavicular brachial plexus injuries?
4. What diagnostic tests would you order?
5. What is the timing for surgery?
6. What types of nerve surgery can be performed?
7. What are the surgical options for delayed (> 1 year) presentation?
8. What are the options for pain management?
9. What other anatomical sites can give the same eye findings?

## Answers

1. *What is the likely diagnosis?*
   - Pan-plexus injury
   - While unlikely, other traumatic injuries that may produce unilateral arm weakness (in addition to other findings) to consider include, but are not limited to:
     – Incomplete cervical spinal cord injury, such as a Brown–Sequard pattern of ipsilateral hemiplegia and contralateral hemianesthesia
     – Traumatic brain injury
     – Vascular dissection with resultant stroke
     – Traumatic cervical disc herniation

## Answers (*continued*)

    - Inflammatory brachial neuritis that causes a patchy plexopathy in a delayed fashion after significant stress. Profound denervation atrophy may occur. A delayed onset and report of severe neuropathic pain are hallmarks of this disease.

2. *What is the impact of the finding of anisocoria on your assessment?*
   - The presence of anisocoria signifies that the T1 nerve root is most likely avulsed.
   - However, there is usually transient anisocoria in patients with T1 injury without avulsion that may produce a similar clinical picture.[1]
3. *What are the common injury patterns in supraclavicular brachial plexus injuries?*
   - Stretch (traction) injury[2]
     - Traction injuries may produce graded injury, from apraxia (transient injury) to axonotmesis (axon injury, which may or may not recover spontaneously) to neurotmesis (nerve discontinuity). There are two types of nerve discontinuities: avulsion (preganglionic injury) and rupture (postganglionic injury) of the involved nerve.
       - ○ Avulsion injuries are typically accompanied by severe, episodic neuropathic pain that is frequently refractory to multiple medications. Patients often provide vivid terms when describing avulsion pain.
       - ○ Avulsion injuries in infants, as occurs in so-called obstetrical brachial plexus palsy (or birth-related brachial plexus palsies), generally do not produce pain-related behaviors.
     - Supraclavicular injuries can be classified into three groups based on the nerve roots involved:
       - ○ Complete brachial plexus injury (C5–T1) (57%)[1]
         - ♦ This injury can lead to total paralysis of the upper extremity or "flail arm."
         - ♦ This injury is associated with highest severity of trauma and, consequentially, the lowest rate of spontaneous recovery.
       - ○ Upper roots injury (Erb's palsy) involving C5–C6 (15%)[2]
         - ♦ This injury can lead to weakness or paralysis of the shoulder (glenohumeral movement, not scapulothoracic), elbow flexion, and pronator/supination movement.
       - ○ Extended upper plexus injury involving C5–C7 (20%)[2]
         - ♦ This injury can lead to weakness or paralysis of the shoulder, elbow flexion, but also includes elbow extension and impairment of movement at the wrist.
       - ○ Lower trunk injury (Klumpke's palsy) involving C8–T1 and C7–T1 (8%)[2]
         - ♦ These injuries can lead to loss of small muscles of the hand resulting in "claw hand," in which loss of lumbrical muscles causes hyperextension of the metacarpophalangeal joint and flexion of the proximal and distal interphalangeal joints.
4. *What diagnostic tests would you order?*
   - MRI of the brachial plexus[1,3–6] provides optimal visualization of the brachial plexus and surrounding soft tissue (▶**Fig. 141.1**).
     - May depict traumatic neuromas as well as presence or absence of nerve continuity.
     - Stretch injury may appear as a thickened or hyperintense plexus on T2-weighted images.
     - Frequently demonstrates denervation changes within muscles, which present early and persist for months (denervated atrophic muscles appear hyperintense on T2-weighted images).
     - Common findings of nerve root avulsion injury on MRI include[7]:
       - ○ Pseudomeningocele, depicted on T-2 weighted images
       - ○ Cord shift from the midline
       - ○ Rootlets are well visualized on coronal fast spin echo sequences (e.g., SPACE, FIESTA, and CISS).
   - CT myelography[1,4] (▶**Fig. 141.1**)
     - The gold standard test for definitive identification of nerve root avulsion (positive finding of injury) and depiction of the continuity of the ventral and dorsal rootlets (negative finding of injury)
     - It has a sensitivity of 92.2% for detecting root avulsion and false positive and false negative rates of 5 to 10%.
     - In case of root avulsion of the lower trunk, the dural sheath often heals with formation of meningocele.
     - CT myelography increases in sensitivity 3 to 4 weeks after the injury, as the blood clot in the injured area dissolves and the meningocele forms.
   - Electrodiagnostic studies, including nerve conduction and EMG[1,4]
     - Nerve conduction studies assess both myelination and axon health by the characteristic conduction properties.
       - ○ Conduction block (neurapraxic injury) demonstrates absent conduction from stimulation proximal to an injury site, but intact stimulation distal to the injury.
       - ○ Demyelination demonstrates slowing velocity, low amplitudes (from temporal dispersion of the peak), and increased latency.
       - ○ Severe axonopathies demonstrate loss of conduction throughout the nerve (although the nerve will often stimulate up to 3 days after injury).
         - ♦ In preganglionic lesions, sensory nerve action potentials are preserved, as the dorsal root ganglion is avulsed with the nerve and the axons remain relatively preserved and myeli-

## Answers (*continued*)

nated. Motor nerve action potentials, however, will be absent.

      - In postganglionic injury, both motor and sensory nerve action potentials will be absent.
    - EMG depicts acute changes due to denervation, i.e., insertional activity, and the response to nerve injury, i.e., recruitment patterns (how much cortical effort), complex motor units, nascent motor units, which may predict spontaneous (or natural) recovery.
      - Insertional activity (e.g., fibrillation potentials) reflects acute denervation of a muscle and is graded subjectively by the EMG diagnostician. Serial EMGs, for observation of an injury for possible recovery, should be performed by the same individual.
      - These can be detected in 10 to 14 days after injury in proximal muscles and in 3 to 6 weeks in distal muscles.
  - X-rays[1,4]
    - Chest
      - Assessment for rib fractures
      - Assess the activity of the diaphragm, as indicator of phrenic nerve involvement. Chest X-ray must be performed at maximal inspiration (during phrenic activation).
    - Cervical spine
      - May demonstrate transverse process fracture indicating possible root avulsion
    - Scapula, shoulder, and clavicle
      - Scapulothoracic dissociation can be seen with root avulsion and vascular injury.
      - Clavicular and humerus fracture may be associated with brachial plexus injury.

5. *What is the timing for surgery?*
   - Immediate surgical exploration[8,9]
     - Should be considered for patients with progressive neurologic decline, vascular injuries, expanding hematoma, open injuries, penetrating trauma, and iatrogenic injury.
     - Gunshot wounds are unique, in that the energy dispersion of the bullet often causes neurapraxia. Exploration should be considered for lesions with high risk of nerve transection, such as military-grade rounds.
   - Within 4 weeks of injury[8,9]
     - Avulsion
       - These patients are candidates for distal nerve transfer within the first 4 weeks of injury.
       - Spinal accessory to suprascapular nerve transfer has been used with good outcomes in complete brachial plexus palsy.[10]
       - Other nerves could be also utilized including medial pectoral nerve branches, thoracodorsal nerve, and intercostal nerves.
     - Some authors advocate for early surgical exploration in cases of clear nerve rupture.[11]
   - Within 3 months of injury[8,9]
     - Stretch injuries
       - Nerve stretch injuries that are not avulsed or clearly ruptured should be evaluated for spontaneous recovery, generally after 3 months, or enough time for remyelination and axon regrowth to the first muscle after the site of the injury.
       - Serial EMGs should be used for observation of nerve lesions, as described above.
     - Low-velocity gunshot wounds
6. *What types of nerve surgery can be performed?*
   - Direct nerve repair[8,12]
     - Best treatment option for acute, sharp penetrating injuries.
     - In traction injuries, end-to-end repair is rarely feasible, because of the long zones of injury and frequent retraction of ruptured nerve endings.
   - Neurolysis[8,12]
     - Performed when low-amplitude regenerative nerve action potentials are produced during intraoperative electrical stimulation.
     - External neurolysis: the fibrous scar surrounding and constricting the injured nerve is dissected off from the nerve.
     - Internal neurolysis: the fibrous scar is dissected off from between the injured nerve fascicles.
     - Generally effective in cases of low-velocity stretch injuries, for example, humerus fractures associated with falls.
   - Nerve grafting[8,12] (▶ **Fig. 141.2**)
     - Performed in cases of postganglionic injury with intact proximal and distal nerve stumps that cannot be directly repaired, or after resection of a nonconducting injured nerve.
     - The sural nerve, medial brachial nerve, and medial antebrachial cutaneous nerve are commonly utilized as nerve grafts.
   - Nerve transfers (neurotization)[8,12,13]
     - Involves transferring a working but less important motor nerve to a nonfunctioning more important denervated muscle.
     - Can be divided into two groups, depending on the source of the transferred nerve:
       - Extraplexal source includes the spinal accessory nerve, intercostal nerves, contralateral C7 nerve, and hypoglossal nerve (▶ **Fig. 141.3**).
       - Intraplexal source includes the phrenic nerve, pectoral nerve, and segment of the median or ulnar nerves.
7. *What are the surgical options for delayed (> 1 year) presentation?*
   - Functioning muscle transfer[12]
     - Gracilis muscle, latissimus dorsi, and others may be as pedicled (one free end) or free muscle transfers (two transferred ends)

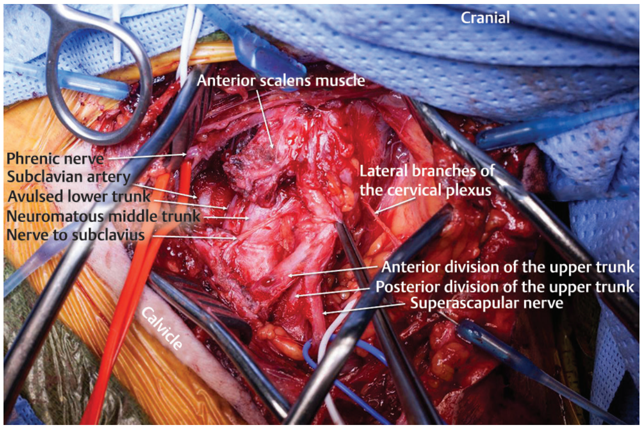
Cranial
Anterior scalens muscle
Phrenic nerve
Subclavian artery
Avulsed lower trunk
Neuromatous middle trunk
Nerve to subclavius
Lateral branches of the cervical plexus
Anterior division of the upper trunk
Posterior division of the upper trunk
Superascapular nerve
Calvicle

**Fig. 141.2** Intraoperative photo of the supraclavicular exploration with anatomical depiction. The C5 and C6 foramen were exposed and no neural elements were found. Shoulder and elbow flexion function would be reconstructed by use of extraplexal sources, including spinal accessory nerve to suprascapular nerve and multiple intercostals to the branch of biceps and musculocutaneous nerve (see ▶**Fig. 141.3**). A severely scarred middle trunk was traced to the C7 foramen and the C7 spinal nerve was found to be viable for transfer or repair. It was transferred to the lower trunk, for the goal of restoring hand function. In addition, three intercostals were also routed to a banked sural graft, for possible future use of a free-functioning muscle transfer to reestablish finger grasp.

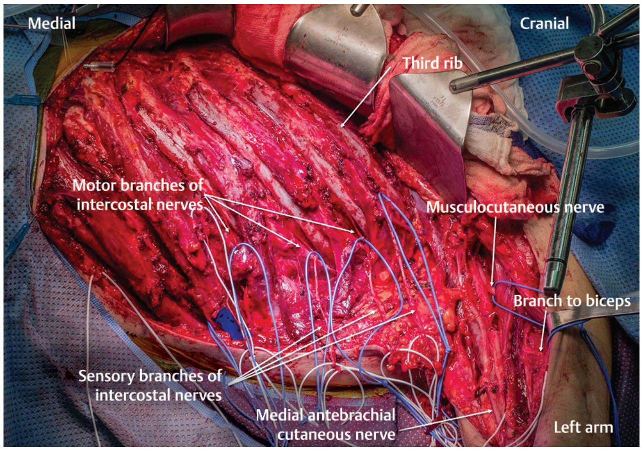
Medial
Cranial
Third rib
Motor branches of intercostal nerves
Musculocutaneous nerve
Branch to biceps
Sensory branches of intercostal nerves
Medial antebrachial cutaneous nerve
Left arm

**Fig. 141.3** Intraoperative photo of the use of intercostal nerves for reanimating elbow flexion. Six intercostal motor nerves were harvested, from beneath the third through the eighth ribs (identified by ***blue*** vessel loops, except ***white*** at R8). Two intercostal motor nerves were transferred directly to the branch to the biceps brachii muscle and one to the brachialis branch of the musculocutaneous nerve (after neurolysis of the musculocutaneous nerve). Lateral sensory branches from the intercostal nerves (identified by ***white*** vessel loops not under the ribs) were sutured to the medial antebrachial cutaneous nerve.

## ■ Answers (*continued*)

  - The results are generally good, but these are performed in highly specialized centers.
- Tendon transfer[14]
  - Best suited for single nerve lesions, such as the Starr set of transfers for radial palsy.
  - Both passive (tendon fixation) and active (tendon rerouting) options exist, providing for function in either the presence or the absence of suitably expendable muscles.

8. *What are the options for pain management?*
   - Medications[15]
     - Anticonvulsants:
       - ○ Gabapentin
         - ♦ 100 to 300 mg at bedtime or 100 to 300 mg three times daily.
         - ♦ Increase by 100 to 300 mg to achieve a therapeutic dose of up to 3,600 mg daily. (Some patients may find benefit below this dose; however, failure should not be assumed until the dose of 3,600 mg daily is achieved).
       - ○ Pregabalin
         - ♦ 50 mg three times daily or 75 mg twice daily.
         - ♦ Increase to 300 mg daily after 3 to 7 days and then by 150 mg daily to a maximum of 600 mg daily.
     - Selective serotonin and norepinephrine reuptake inhibitors:
       - ○ Duloxetine
         - ♦ 30 mg once daily.
         - ♦ Increase to 60 mg once daily to a maximum of 60 mg twice daily.
         - ♦ Should be taken for 4 weeks.
       - ○ Venlafaxine
         - ♦ 37.5 mg once or twice daily.
         - ♦ Increase by 75 mg each week to a maximum of 225 mg daily.

## Answers (*continued*)

        - Should be taken for 4 to 6 weeks.
  - Tricyclic antidepressants:
    - Nortriptyline and desipramine
      - 25 mg at bedtime.
      - Increased by 25 mg to a maximum of 150 mg daily.
      - Should be taken for 6 to 8 weeks.
  - Five percent lidocaine patch
    - Maximum three patches daily for a maximum of 12 hours.
    - Should be applied for 3 weeks.
  - Opioids
    - Morphine, methadone, and oxycodone
      - Second-line medication
      - 10 to 15 mg every 4 hours or when necessary
      - Should be taken for 4 to 6 weeks
  - Tramadol
    - Second-line medication
    - 50 mg once or twice daily
    - Increase by 50 to 100 mg daily to a maximum of 400 mg daily
    - Should be taken for 4 weeks.
- DREZotomy[16–18]
  - Dorsal Root Entry Zone (DREZ) lesioning is the ablation of the lateral portion of the dorsal rootlets, hyperactive neurons of dorsal horn, and excitatory portion of Lissauer's tract for control of pain.
  - Seventy-five to 98.2% of patients with intractable pain from brachial plexus avulsion report good results immediately after surgery.[16–18]
  - However, the pain relief rate decreases gradually during long-term follow-up.
  - In one study, immediate results provided good pain relief (i.e., > 75% pain relief) in 79% of patients, and long-lasting good pain relief in 66.5% of patients with 5% morbidity rate.[16]
- Cervical spinal cord stimulation[19]
  - Cervical spinal cord stimulation or dorsal column stimulation for brachial plexus avulsion intractable pain achieves pain relief in 43 to 100% of patients.

9. *What other anatomical sites can give the same eye findings?*
   - There are three types of injury that can produce Horner's syndrome, depending on the level of the injured neurons[20–22]:
     - First-order neuron lesions
       - These may present with hemisensory loss, dysarthria, dysphagia, ataxia, vertigo, and nystagmus.
       - Anhidrosis affects the ipsilateral side of the body.
       - Causes include basal meningitis or skull base tumors, pituitary tumors, Chiari malformations, syringomyelia,[23] multiple sclerosis, stroke with Wallenberg syndrome (lateral medullary syndrome), neck trauma, and/or dissection of the vertebral artery.
     - Second-order neuron lesions
       - These may present with facial, neck, axillary, shoulder or arm pain, cough, hemoptysis, history of thoracic or neck procedures or trauma, or anhidrosis of the ipsilateral face.
       - Causes include birth trauma, Pancoast tumor or cervical rib, central venous line placement, chest tube placement,[24] aortic dissection, hilar lymphadenopathy, trauma or surgical injury to the neck or upper thorax, or middle ear lesions.[25]
     - Third-order neuron lesions
       - These may present with diplopia from sixth nerve palsy, numbness in the distribution of the first or second division of the trigeminal nerve, and pain.
       - Anhidrosis is either absent or limited (fibers travel with the external carotid artery).
       - Causes include carotid cavernous fistula, internal carotid artery dissection,[26] herpes zoster, and Raeder's paratrigeminal syndrome.
   - See ▶ **Fig. 141.4** for a summary of Horner's syndrome pathways.

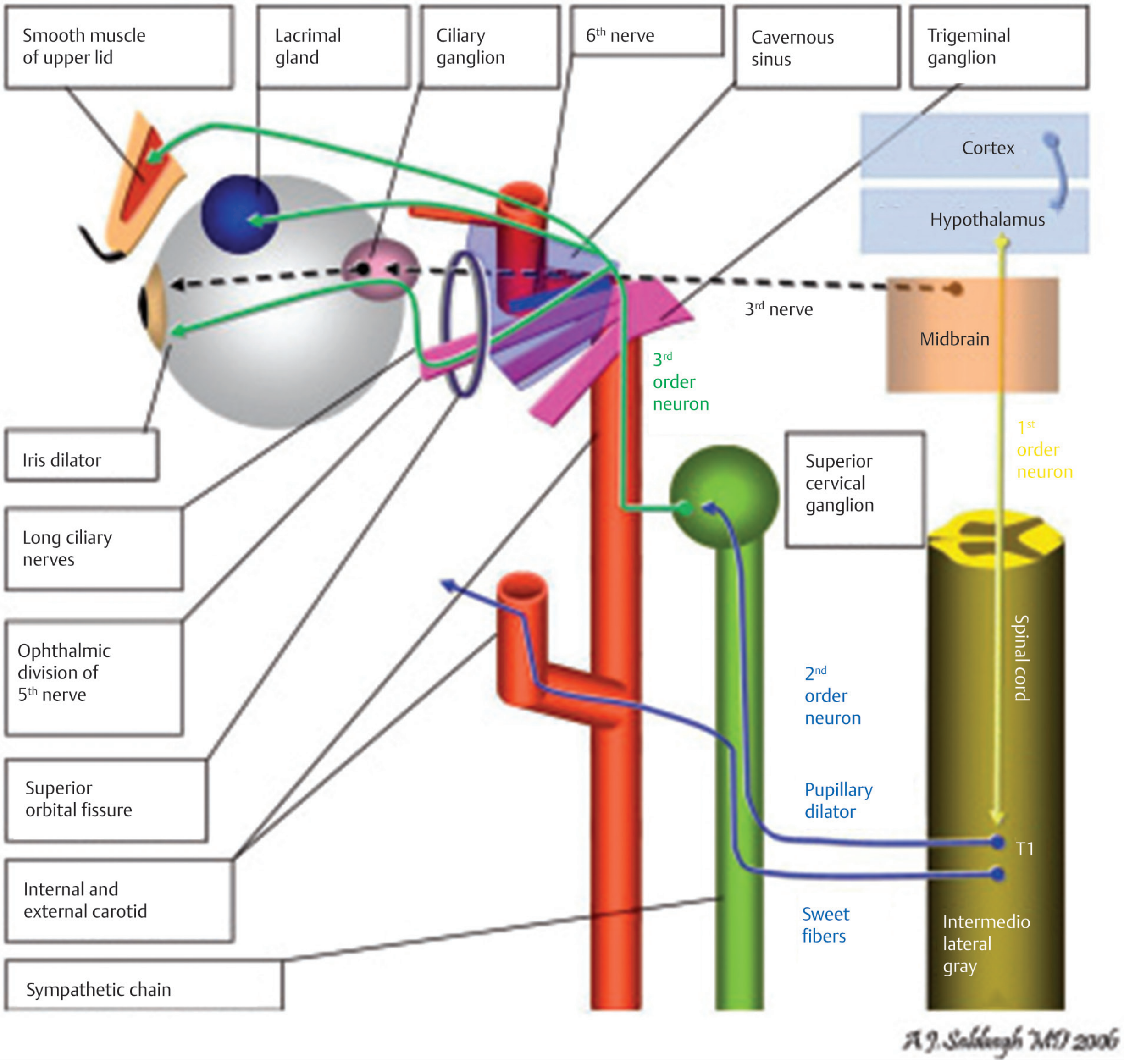
Smooth muscle of upper lid
Lacrimal gland
Ciliary ganglion
6th nerve
Cavernous sinus
Trigeminal ganglion
Cortex
Hypothalamus
3rd nerve
Midbrain
3rd order neuron
1st order neuron
Iris dilator
Long ciliary nerves
Superior cervical ganglion
Spinal cord
Ophthalmic division of 5th nerve
2nd order neuron
Superior orbital fissure
Pupillary dilator
T1
Internal and external carotid
Sweet fibers
Intermedio lateral gray
Sympathetic chain
A.J. Sabbagh MD 2006

**Fig. 141.4** Horner's syndrome—anatomic depiction of the involved pathways. (**a**) First-order neurons (central sympathetic) start at the posterolateral hypothalamus, descend uncrossed via the midbrain and pons, and end in the intermediolateral gray at the level of C8 to T2. (**b**) Second-order preganglionic pupillomotor fibers leave the spinal cord at T1 and join the cervical sympathetic chain close to the pulmonary apex. These nerves ascend through the sympathetic chain and synapse in the superior cervical ganglion near the bifurcation of the common carotid artery (around C3 to C4 levels). (**c**) Third-order neurons: pupillomotor fibers (postganglionics) leave the superior cervical ganglion and migrate along the internal carotid artery. Vasomotor and the sudomotor fibers branch off shortly after the postganglionic fibers leave the superior cervical ganglion. These fibers travel along the external carotid artery to innervate the blood vessels and sweat glands of the face. The pupillomotor fibers enter the cavernous sinus while traveling along the internal carotid artery. These fibers then join the abducens nerve (CN VI) in the cavernous sinus and enter through the superior orbital fissure into the orbit along with the ophthalmic branch of the trigeminal nerve (CN V1) via the long ciliary nerves. The long ciliary nerves innervate Müller muscle and the iris dilators. (Reproduced from Patten JP. Neurological Differential Diagnosis. 2nd ed. Glasgow: Springer Verlag; 1996. Reede DL, Garcon E, Smoker WR, Kardon R. Horner's syndrome: clinical and radiographic evaluation. Neuroimaging Clin N Am 2008;18(2):369–385 xi. Bardorf CM. Horner syndrome. emedicine.com 2006. Available at: http://www. emedicine.com/OPH/topic336.htm. Accessed April 22, 2009. Nader R., et al. Neurosurgery Case Review. New York: Thieme; 2010.)

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# Case 142 Median Nerve Laceration at the Wrist (Spaghetti Wrist)

*Bassam M. J. Addas*

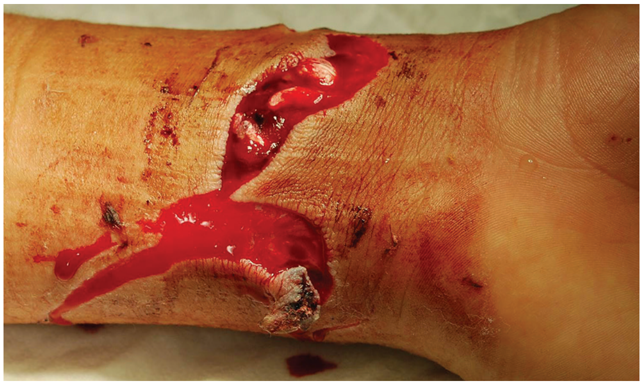

**Fig. 142.1** Preoperative photograph depicting glass laceration of the left wrist.

## Clinical Presentation

- A 25-year-old male was injured by running into a glass door and presents to emergency room with his left wrist wrapped in his own shirt to control bleeding.
- Following vital signs assessment the distal wrist was exposed to the reveal a ragged laceration with no excessive active bleeding (▶**Fig. 142.1**).
- The hand was pale, cold, and insensate in the territory of both median and ulnar nerves distribution and the pulses in the palmar arch were not palpable. No wrist or finger flexion was possible.
- Sensation at the dorsum of the hand was intact and finger extension was possible.

## Questions

1. What is the common term used to describe this type of injury?
2. What are the common causes of “spaghetti wrist” injury?
3. How can you differentiate between glass injury and suicide attempt?
4. What is the emergency management of such an injury?
5. When do these injuries need to be repaired?
6. What are the commonly injured structures in spaghetti wrist injuries?
7. What are the instruments needed in the repair of the distal median and ulnar nerves?
8. What are the priorities in the repair of such injuries?
9. What is the postoperative care plan?

## Answers

1. *What is the common term used to describe this type of injury?*
   - “Spaghetti wrist” is a term used to describe such an injury where there is injury to at least three or more structures of the distal forearm including one nerve or one artery at least. It likens it to spaghetti because the injured elements (usually nerves and tendons) are somehow similar to spaghetti in shape and consistency.[1–3]
2. *What are the common causes of “spaghetti wrist” injury?*
   - Glass injuries, altercations involving sharp objects, and occasionally suicide attempts.[3]
3. *How can you differentiate between glass injury and suicide attempt?*
   - When confronted with sharp lacerations of the distal wrist, it is important to differentiate between glass injury and suicide attempts for legal and counselling considerations.
   - Glass injury can involve the dominant and the nondominant hand whereas the suicide attempts usually involves the nondominant hand only.
   - Suicide attempt usually produce one sharp nonragged laceration, whereas accidental glass laceration produces multiple, angled wounds with variable depths.

## Answers (*continued*)

4. *What is the emergency management of such an injury?*
    - ABCDE of trauma should be cleared first (securing airway, breathing, systemic circulation, general disability assessment, and exposure of any injured parts of the body).
    - In the emergency department, control of bleeding with attention to blood pressure and administration of tetanus toxoid are most important.
    - The wound is then covered and preparation for surgery is initiated.
    - Perioperative intravenous (IV) antibiotics are administered.
    - Care should be exercised while washing the wound in the emergency room as it may dislodge clots and reinitiate bleeding. This should be rather performed in the operating theatre with the patient anesthetized.
5. *When do these injuries need to be repaired?*
    - The best chance for maximizing the patient's recovery is to repair these injuries in the acute setting.[2]
    - If the patient presents in the late hours of the day, it is reasonable to book them the next morning to assure the most optimal operating room team and environment.
    - Acute nerve lacerations are addressed optimally via primary repair with the best results obtained in this fashion.
    - Delay of repair will cause retraction of the nerves and make their primary repair difficult with the possible need of grafts which decreases the percentage of good results.
6. *What are the commonly injured structures in spaghetti wrist injuries?*
    - Wrist and finger flexor tendons, median and ulnar nerves, radial and ulnar arteries[3] occasionally accompanied by distal radius and ulna fractures (▶**Fig. 142.2**).
    - See ▶**Fig. 142.3** for illustration.
7. *What are the instruments needed in the repair of the distal median and ulnar nerves?*
    - Optimal loupe magnification (4.5×) using 8/0 nylon sutures will allow adequate epineurial sutures to be placed efficiently. Larger size sutures and lack of magnifications will cause more injuries to the nerve making it difficult to achieve a finer epineurial approximation.
    - *The use of tissue glue alone without sutures is unacceptable.*
    - Microinstruments are available in most neurosurgical units and if not they can be borrowed from ophthalmology service in the facility.
    - Microscope, if available, can be used but it is usually not necessary in primary repair of major nerves.[3–5]
8. *What are the priorities in the repair of such injuries?*
    - Vascular injuries carry the priority in the repair to reestablish circulation to the injured hand, followed by the tendon repair, the last structures repaired are the nerves.[3] The nerves are kept to the end to minimize manipulation of the field following their delicate repair as, if performed sooner, may be disrupted and require revision (▶**Fig. 142.4**, ▶**Fig. 142.5**).
9. *What is the postoperative care plan?*
    - The wrist is placed in 15 degree of gentle flexion in a posterior slab for 6 weeks allowing both nerve and tendons to heal.
    - This is followed by rigorous physical therapy.
    - The patient needs to be reminded that nerve recovery may take up to 6 months and may not be complete, and excellent results are only expected in up to 75% of cases.[3,6]

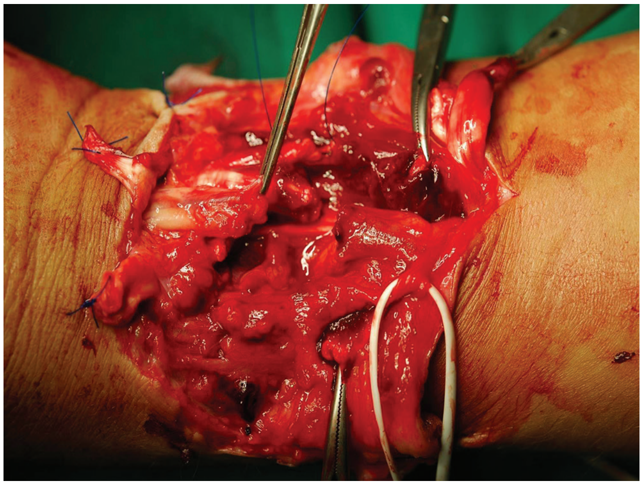

**Fig. 142.2** Intraoperative photograph following the exposure of the wrist and identification of the involved structures, including the flexor tendons, the median and the ulnar nerves, and the radial artery.

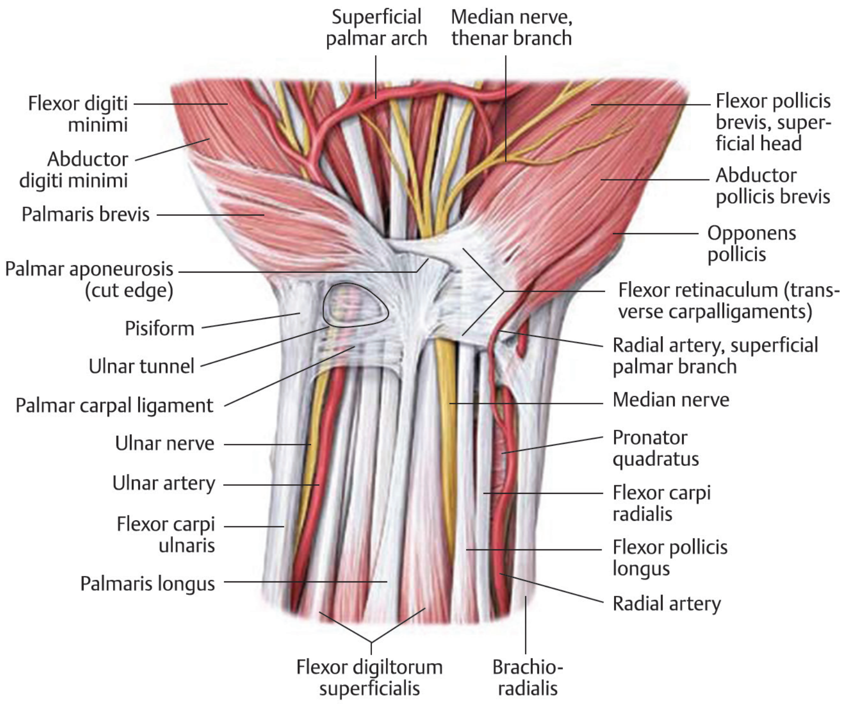
Superficial palmar arch
Median nerve, thenar branch
Flexor digiti minimi
Abductor digiti minimi
Palmaris brevis
Palmar aponeurosis (cut edge)
Pisiform
Ulnar tunnel
Palmar carpal ligament
Ulnar nerve
Ulnar artery
Flexor carpi ulnaris
Palmaris longus
Flexor pollicis brevis, superficial head
Abductor pollicis brevis
Opponens pollicis
Flexor retinaculum (transverse carpalligaments)
Radial artery, superficial palmar branch
Median nerve
Pronator quadratus
Flexor carpi radialis
Flexor pollicis longus
Radial artery
Flexor digiltorum superficialis
Brachioradialis

**Fig. 142.3** Detailed anatomic rendering of the neurovascular structures along the palmar aspect of the wrist. Coursing tendons and muscles are also identified. (Reproduced with permission from Schuenke M. et al. Thieme Atlas General Anatomy and Musculoskeletal System. New York: Thieme; 2010, 357. Illustration by Karl Wesker/Markus Voll.)

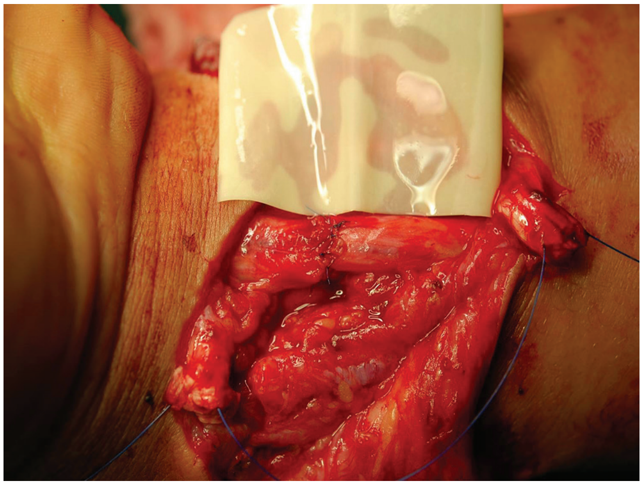

**Fig. 142.4** Following the repair of the radial artery and the flexor tendons, the flexor carpi radialis tendon is isolated and reflected before the median nerve repair.

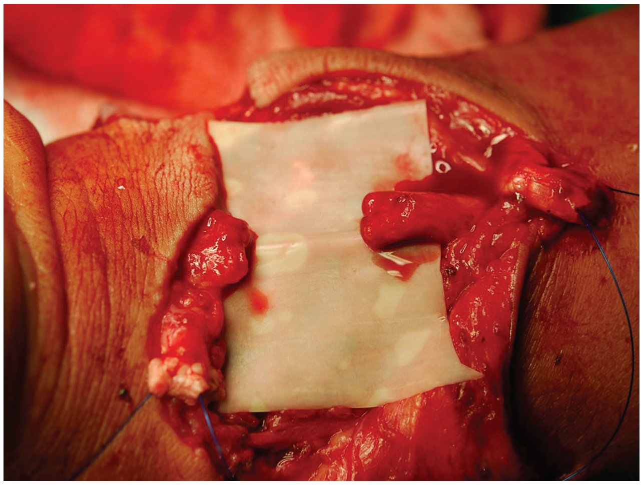

**Fig. 142.5** Following repair of the median nerve with alignment of the fascicles aided by the surface vessel on the nerve. The flexor carpi radialis was not sutured as it may compress the median nerve repair, this situation is not universal and every case requires assessment (▶**Fig. 142.4**). The wrist flexion will still be adequate with flexor carpi ulnaris and palmaris longus repair.

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# Case 143 Radial Nerve Injury

*Frank Gerold and Jaime Gasco*

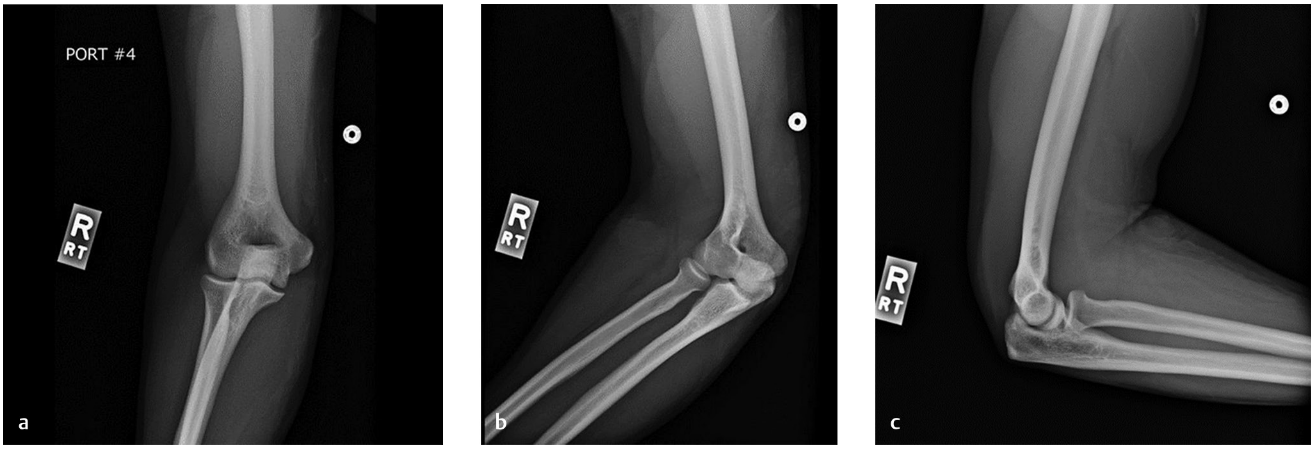

**Fig. 143.1** Elbow X-ray images with frontal (**a**), condyle (**b**), and lateral views, showing no obvious fractures or dislocations (**c**).

## Clinical Presentation

- A 23-year-old right-hand-dominant man presents after a right forearm laceration and hand weakness after punching through a pane of glass.
- The patient states that he feels weakness in his hand and has difficulty extending the fingers and thumb. He also complains of altered sensation on the dorsal radial aspect of his hand.
- Examination of the right upper extremity reveals a transverse laceration over the dorsoradial aspect of his forearm just distal to the antecubital fossa.
  - He can actively flex and extend the elbow without difficulty.
  - There is radial deviation with attempted wrist extension. He cannot extend his fingers and cannot extend his thumb.
  - His wrist flexors and finger flexors are intact. Hand intrinsics are intact.
  - Sensation is intact in the median and ulnar nerve distribution; however, it is altered in the radial nerve distribution.
- X-rays of the right elbow show no foreign body or obvious fracture (see ▶**Fig. 143.1**).

## Questions

1. What is the correct diagnosis?
2. Describe the anatomic course of the radial nerve.
3. At what level is the nerve injured in this case?
4. How can one differentiate between a radial nerve proper injury and a posterior interosseous nerve (PIN) injury on physical exam?
5. What are the surgical indications after a radial nerve injury?
6. What are the surgical options for nerve repair?
7. How is the patient managed postoperatively?
8. What is the expected outcome?
9. What is the order of recovery of the muscles innervated by the radial nerve in the forearm?
10. Are there any other surgical options to restore function after a radial nerve injury?

## Answers

1. *What is the correct diagnosis?*
   - Based on the functional deficits, the patient has an injury to his radial nerve.
   - In the forearm, the radial nerve is responsible for motor innervation to the wrist and finger extensors. It supplies sensation to the dorsum of the hand specifically on the radial two-thirds.
   - Distal to the triceps, the radial nerve proper innervates the brachioradialis (BR), the extensor carpi radialis longus (ECRL), and the anconeus muscles.
   - The extensor carpi radialis brevis (ECRB) and supinator are innervated by the deep branch of the radial nerve.
     - Of note, the ECRB is known to receive variable innervation from the radial nerve proper, PIN, and even the superficial branch of the radial nerve (SBRN).[1]
   - The remainder of the muscles in the posterior compartment of the forearm are innervated by the PIN which is the continuation of the deep branch after it passes through the supinator muscle.
2. *Describe the anatomic course of the radial nerve.*
   - The radial nerve can be identified deep to the axillary artery as the terminal branch of the posterior cord of the brachial plexus.
   - It courses through the triangular interval bordered by the teres major, long head of the triceps, and humeral shaft.
   - The nerve then wraps around the spiral groove in a proximal medial to distal lateral direction at an average 14 cm proximal to the lateral epicondyle.[2]
   - It pierces the lateral intermuscular septum 10 cm proximal the lateral epicondyle and enters the anterior compartment emerging between the brachialis and BR muscles.[2]
   - It then divides into the deep branch and SBRN approximately 1 cm proximal to the radiocapitellar joint.[3]
   - The deep branch pierces the supinator and is named the PIN after it emerges and runs on the dorsal aspect of the interosseous membrane, giving off motor branches, until it terminates into the dorsal wrist capsule via the floor of the fourth dorsal extensor compartment.
   - The SBRN courses with the radial artery in the forearm deep to the BR which emerges superficially 8 cm proximal to the radial styloid to provide sensation to the dorsum of the hand.[4]
   - See ▶**Fig. 143.2** for illustrated course.
3. *At what level is the nerve injured in this case?*
   - The patent is showing signs of preserved ECRL function as demonstrated by radial deviation on attempted wrist extension. However, he cannot extend the wrist or fingers. He also has decreased sensation in the radial nerve distribution in the hand.
   - Therefore, the level of the injury is distal to the radial nerve proper and involves the deep branch of the radial nerve as well as the SBRN.
4. *How can one differentiate between a radial nerve proper injury and a posterior interosseous nerve (PIN) injury on physical exam?*
   - A radial nerve proper injury will present with lack of both wrist and finger extension in addition to a sensory deficit.
   - A PIN injury will present with a pure motor deficit and preserved sensation as the SBRN is intact. It is possible to observe some wrist extension with an innervated ECRL and possibly some ECRB depending on its variable innervation and level of injury.
5. *What are the surgical indications after a radial nerve injury?*
   - If there is a penetrating injury such as a laceration or open fracture, the nerve should be explored.
   - If there is a blunt injury or a closed fracture and the injury is presumed neuropraxic, then observation is warranted. If after 3 months there is no meaningful recovery on physical exam and on electromyography (EMG), then surgery is indicated.
6. *What are the surgical options for nerve repair?*
   - If the nerve injury is acute and from a sharp laceration or penetrating injury, then primary neurorrhaphy may be possible. It is essential to maintain a tension free repair, however.
   - Once the damaged ends are appropriately resected and a gap is present, then reconstruction with autograft versus allograft is indicated.
   - In this case, sural nerve autograft was utilized for the reconstruction (see ▶**Fig. 143.3**, ▶**Fig. 143.4**, and ▶**Fig. 143.5** for details of reconstruction).
7. *How is the patient managed postoperatively?*
   - The patient is splinted with the elbow flexed at 90 degrees to eliminate any tension across the repair.
   - At the first postoperative visit between 7 and 10 days, the patient is sent to the hand therapist for removable splint application and protected elbow range of motion. The splint is quickly weaned to prevent an elbow contracture.
   - It is important to support the wrist and fingers with a cock-up wrist splint and range of motion exercises to prevent joint contractures while the nerve regenerates.
8. *What is the expected outcome?*
   - It is important to counsel patients on the expected outcomes after peripheral nerve injuries as it can take many months to see recovery.
   - Peripheral nerves regenerate on average 1 mm per day.[5]
   - Nerve injuries are time sensitive matters and yield superior results if addressed early rather than in a

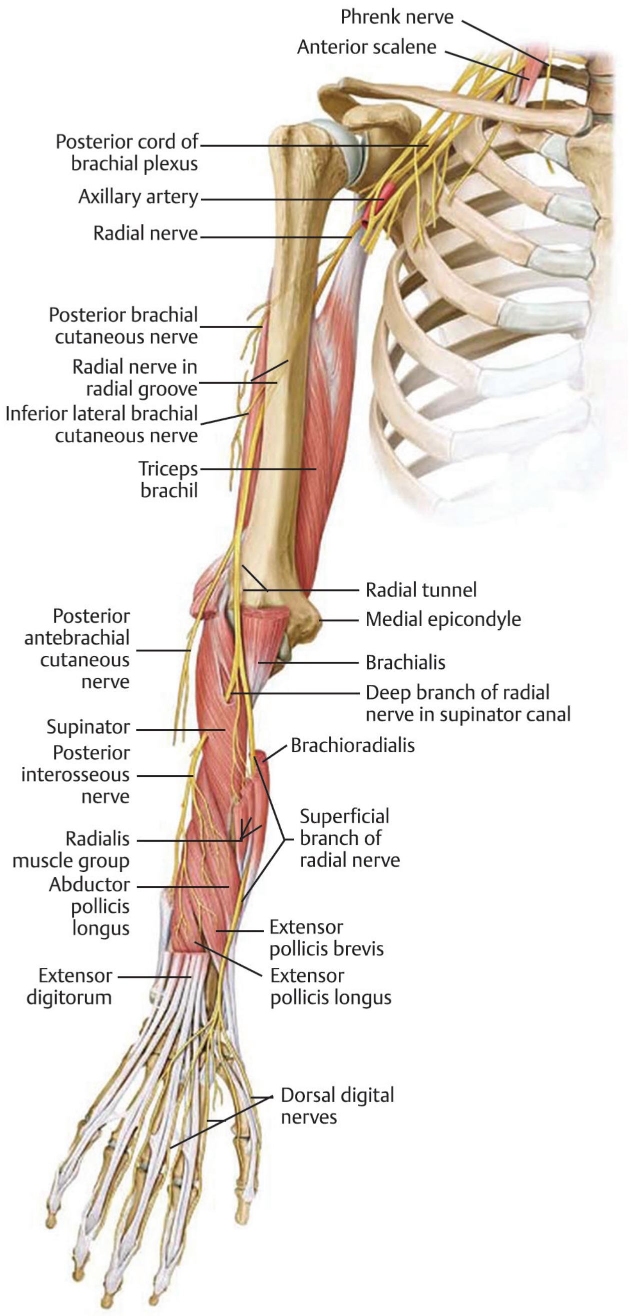
Phrenk nerve
Anterior scalene
Posterior cord of brachial plexus
Axillary artery
Radial nerve
Posterior brachial cutaneous nerve
Radial nerve in radial groove
Inferior lateral brachial cutaneous nerve
Triceps brachil
Radial tunnel
Medial epicondyle
Posterior antebrachial cutaneous nerve
Brachialis
Deep branch of radial nerve in supinator canal
Supinator
Brachioradialis
Posterior interosseous nerve
Superficial branch of radial nerve
Radialis muscle group
Abductor pollicis longus
Extensor pollicis brevis
Extensor digitorum
Extensor pollicis longus
Dorsal digital nerves

**Fig. 143.2** Detailed anatomic rendering of the course of the radial nerve from its origin along the posterior cord of the brachial plexus. Innervated muscles are also identified and labeled. (Reproduced with permission from Schuenke M. et al. Thieme Atlas General Anatomy and Musculoskeletal System. New York: Thieme; 2010, 323. Illustration by Karl Wesker/Markus Voll.)

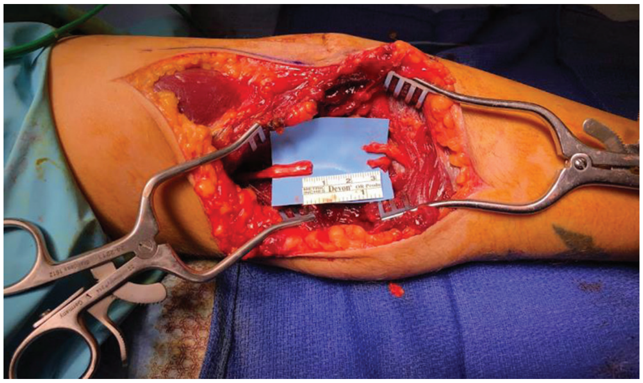

**Fig. 143.3** Intraoperative photograph following the exposure of the radial aspect of the elbow and forearm with identification of the involved structures, including the radial nerve and distal branches (posterior interosseous nerve [PIN], and superficial branch of the radial nerve [SBRN]). A 2.5 cm gap is appreciated between the proximal and distal branches, after damaged ends have been appropriately resected.

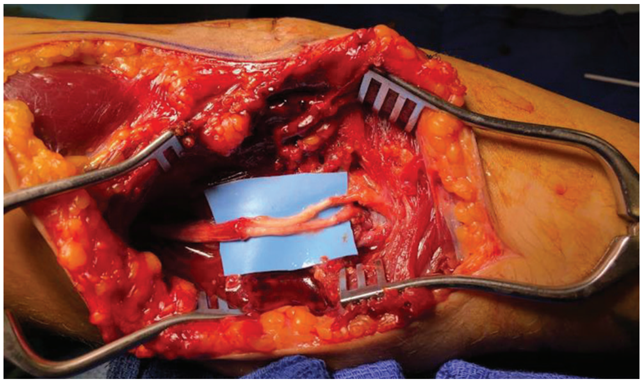

**Fig. 143.4** Intraoperative photograph following repair of the radial nerve with alignment of the fascicles aided by surface vessel on the nerve. Sural nerve autograft was used for fascicular repair.

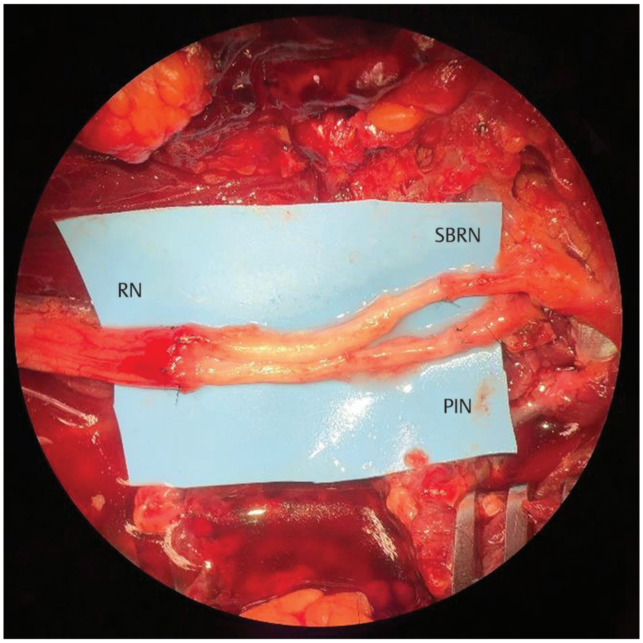

**Fig. 143.5** Intraoperative photograph showing close-up of repair of the radial nerve and continuity with posterior interosseous nerve and superficial branch of the radial nerve.

## Answers (*continued*)

delayed fashion. Other factors such as patient age and graft length may also affect the outcome.[6]

- Radial nerve repair and grafting can result in meaningful motor and sensory recovery with approximately 80% of patients seeing grade 3 recovery most notably in wrist extension.[6,7]

9. *What is the order of recovery of the muscles innervated by the radial nerve in the forearm?*
   - From proximal to distal the order of recovery is as follows[8]:
     - BR
     - ECRL
     - Supinator
     - ECRB
     - Extensor digitorum communis (EDC)
     - Extensor carpi ulnaris (ECU)
     - Extensor digiti minimi (EDM)
     - Abductor pollicis longus (APL)
     - Extensor pollicis longus (EPL)
     - Extensor pollicis brevis (EPB)
     - Extensor indicis proprius (EIP)
   - It is important to know the order of innervation when monitoring for recovery postoperatively. The first muscle to return is BR and the last is EIP.
10. *Are there any other surgical options to restore function after a radial nerve injury?*
    - Tendon transfers have traditionally been utilized to restore wrist and finger extension.[9]
    - Recently, a median to radial nerve transfer has been described and shown to be successful in treating radial nerve injuries.[10]

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# Case 144 Axillary Mass

*Brian Gill and Christopher J. Winfree*

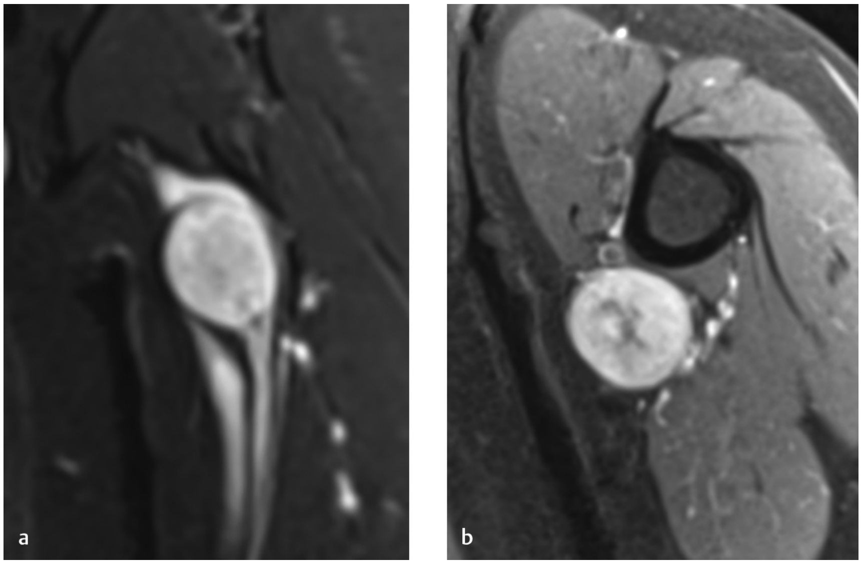

**Fig. 144.1** Coronal T2 MRI (**a**) and axillary T1 contrast-enhanced MRI (**b**) of an axillary schwannoma, demonstrating a well-circumscribed, benign lesion with no edema or contrast enhancement tracking into the surrounding tissues. Also note the relationship of the tumor to surrounding vascular structures.

## Clinical Presentation

- A 47-year-old right-handed man, with no significant past medical history, presents with complaints of a palpable mass in his right axilla for 1 year. He denies any change in the size of the mass over this time. He also denies the presence of pain, sensory abnormalities, or weakness in the involved extremity.
- Peripheral nerve examination of the involved extremity revealed no sensory deficits and full strength in all muscle groups.
- Percussion of the mass elicits a tingling sensation along the medial aspect of the arm, forearm, and the fourth and fifth digits.

## Questions

1. What is the differential diagnosis for this patient?
2. What components of the history and physical examination suggest a benign versus malignant diagnosis?
3. What additional tests should be ordered?
4. What is the appropriate course of treatment?
5. What is the role for radiotherapy in the management of axillary masses?
6. What are some of the prognostic factors for patients with malignant peripheral nerve sheath tumor (MPNST)?

## Answers

1. *What is the differential diagnosis for this patient?*
    - The case presentation describes an otherwise asymptomatic axillary mass with a positive Tinel's sign on palpation. The differential for this may include benign and malignant primary nerve sheath tumors, and a variety of peripheral nonneural sheath plexus tumors (PNNST).[1,2]
    - Benign nerve sheath tumors such as schwannomas and neurofibromas are the most common axillary region tumors, with the latter being more prevalent

## Answers (*continued*)

in most published series. MPNSTs including malignant schwannomas and neurofibrosarcomas are also a possibility.[1]

- PNNSTs are a group of tumors and tumor-like conditions including connective tissue lesions such as lipomas, sarcomas, and desmoid tumors as well as metastatic lesions.[2] These lesions may generate a Tinel's sign due to extrinsic compression of nearby nerves.

2. *What components of the history and physical examination suggest a benign versus malignant diagnosis?*
   - Benign lesions of the axilla typically present as painless, slow-growing masses. In addition, these lesions often have greater mobility in the transverse plane as opposed to the longitudinal plane along the course of the nerve.[3]
   - In contrast, malignant lesions of the axilla are generally painful, firm, fixed masses that display rapid growth and progressive loss of neurologic function.[3]
3. *What additional tests should be ordered?*
   - MRI is the most useful adjunctive test. The various pulse sequences available enable the identification of the location of the mass and its relationship to surrounding neurovascular structures (▶**Fig. 144.1**).[4]
   - The addition of contrast can add more information about the tumor. For example, contrast extravasation into surrounding tissues may suggest a more malignant lesion.
   - MR neurography may be obtained, if available, as it produces greater resolution of the nerves and the surrounding soft-tissue structures.[5]
   - Preoperative electrophysiological studies, such as electromyography and nerve conduction studies, while not critical, may aid in identifying subclinical deficits prior to surgical intervention.[6]
4. *What is the appropriate course of treatment?*
   - Brachial plexus exploration for gross total resection of the lesion is performed in order to obtain a diagnosis, establish prognosis, provide symptomatic relief, and prevent progression of disease.
   - Lesions involving the roots and trunks of the brachial plexus are best approached using an anterior supraclavicular approach. In contrast, the infraclavicular approach is best for lesions that lie in close relation to the cords and distal plexal elements.
   - Benign tumors, such as schwannomas and neurofibromas, are often amenable to gross total resection. The proximal and distal poles of the lesion are identified and it is then separated and mobilized from surrounding structures. In schwannomas most fascicles course outside the capsule of the lesion, with few running through the tumor itself. Conversely, neurofibromas often have several fascicles traversing through the body of the lesion. In either case interfascicular dissection and intraoperative nerve action potential (NAP) studies are performed in order to identify nonfunctioning fascicles that may be removed in order to facilitate tumor removal[1,7] (▶**Fig. 144.2**, ▶**Fig. 144.3**, and ▶**Fig. 144.4**).
   - When there is strong clinical suspicion for a more malignant lesion, positron emission tomography (PET) scanning may be obtained first. This imaging study may suggest the presence of a malignant lesion based upon how "hot" the lesion appears. PET imaging may suggest the optimum location of the lesion to biopsy in the case of a large, malignant lesion, to reduce the likelihood of a nondiagnostic biopsy. Also, PET imaging can suggest the presence of metastatic lesions which would alter the overall management of the patient, and preclude the need for gross total excision of the lesion.
   - If a malignancy is suspected, a needle biopsy can help confirm the diagnosis. Advantages of the needle biopsy compared to open excisional biopsy are the lower risk of tissue plane violation in the setting of malignant lesions which can facilitate subsequent radical en bloc excision. One disadvantage of the needle biopsy is that it is more prone to sampling error than an open excisional biopsy.
   - CT scans of the chest, abdomen, and pelvis should be obtained to evaluate for the presence of a metastasis or a primary lesion prior to the second surgery. The goal of surgery in this setting is gross total resection with wide tumor-free margins.[8] This is often difficult without sacrificing major neurovascular bundles and subtotal resection followed by adjuvant radiation or forequarter amputation in order to achieve oncologic goals.[1]
5. *What is the role for radiotherapy in the management of axillary masses?*
   - Axillary schwannomas and neurofibromas are benign lesions that are usually managed with gross total resection alone.
   - Adjuvant radiotherapy is recommended for MPNSTs as it has been shown to provide local control and delay the onset of recurrence. It may be administered postoperatively or preoperatively with the latter reserved for lesions whose location, size, and distribution make it technically difficult to provide radiotherapy following surgical resection.[9]
6. *What are some of the prognostic factors for patients with malignant peripheral nerve sheath tumor (MPNST)?*
   - Patients with neurofibromatosis type 1 (NF1), surgical margins that contain evidence of tumor, lesions > 5 cm in size, and higher tumor grade have been shown to be poor prognostic factors for patients with MPNST.[9]

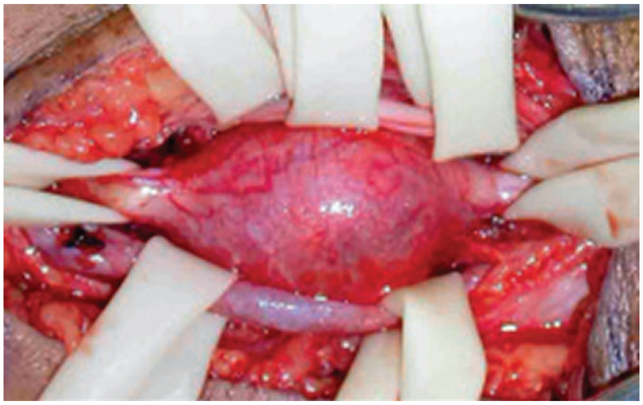

**Fig. 144.2** Intraoperative photograph during infraclavicular brachial plexus exploration. Penrose drains have been used to isolate and retract neurovascular elements. The ulnar nerve, in the center of the field, has undergone fusiform enlargement by the tumor. The median nerve is seen in the upper portion of the exposure, while the basilic vein is seen below.

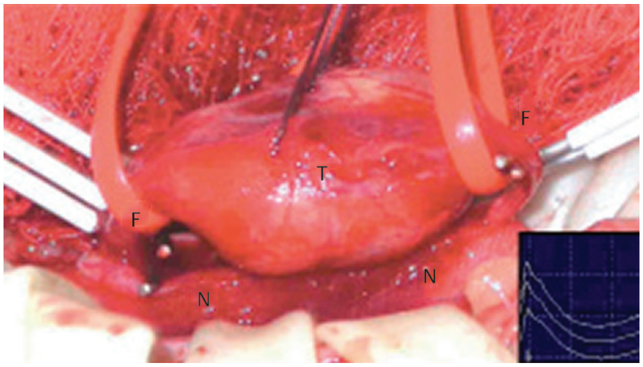

**Fig. 144.3** Intraoperative photograph during excision of the tumor. The tumor (T) has been separated from the parent nerve (N), and remains attached by afferent and efferent fascicles (F). Intraoperative nerve action potential recordings (Inset, flat tracing) using special hook electrodes show that the attached tumor-associated fascicles are nonfunctional and may be sacrificed without yielding a neurologic deficit.

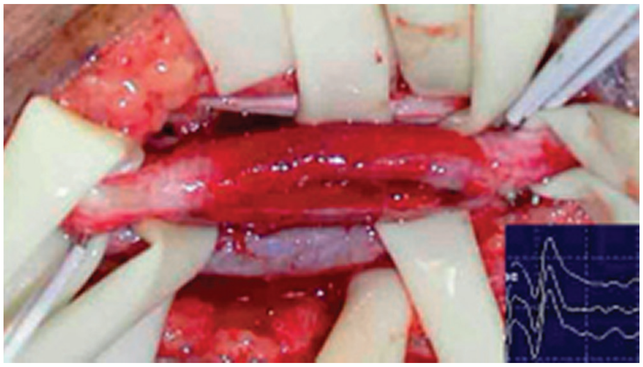

**Fig. 144.4** Intraoperative photograph following gross total excision of the tumor. Intraoperative nerve action potential recordings (Inset, robust tracing) using special hook electrodes show that the ulnar nerve, though bruised, remains both anatomically and functionally intact.

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# Case 145 Medial Arm Mass

*Bassam M. J. Addas*

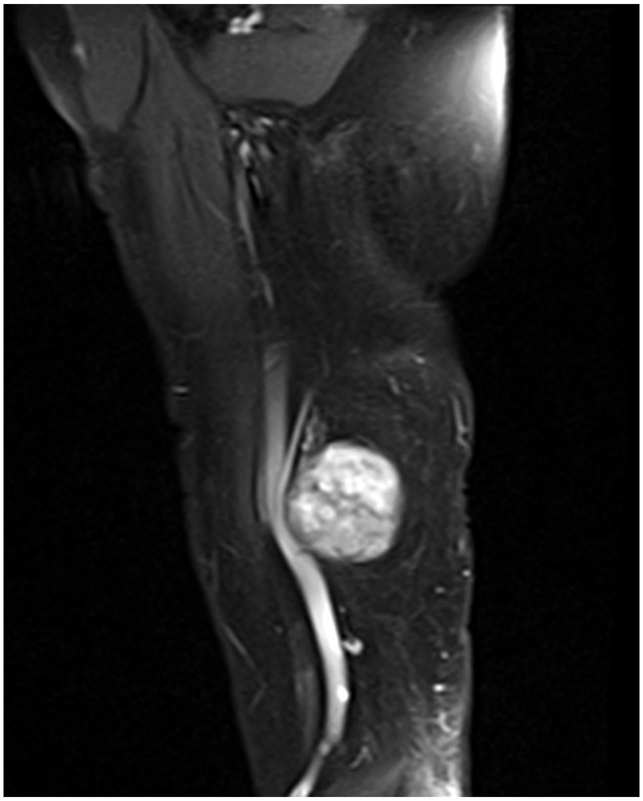

**Fig. 145.1** MRI T1-weighted image, fat saturation with gadolinium contrast of the right arm depicting a round mass with clear borders from the surrounding muscles, with homogenous enhancement following contrast administration. There is no edema detected in the surrounding structures.

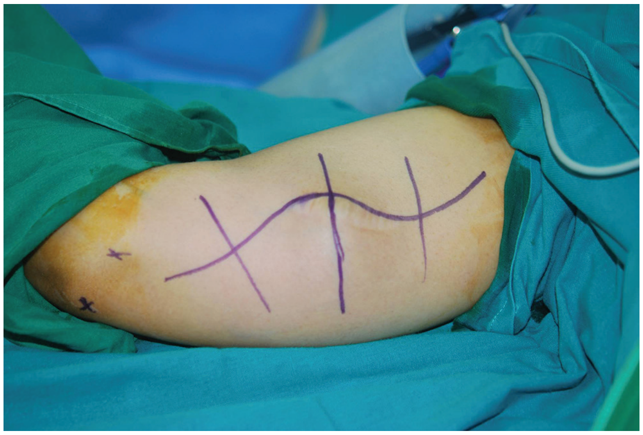

**Fig. 145.2** The planned incision, note the length of the incision compared to the tumor diameter. As a general rule, the incision needs to be double the diameter of the tumor (whenever possible).

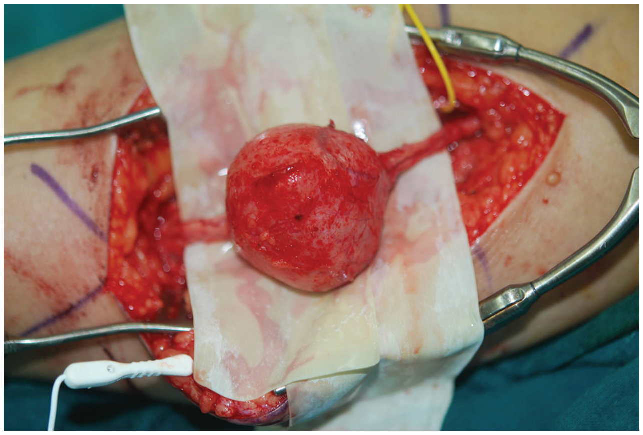

**Fig. 145.3** The ulnar nerve and the tumor following external neurolysis.

## Clinical Presentation

- A 32-year-old woman presents with a right medial arm mass that has been present for 7 years. The patient went for surgery 5 years ago at a different institute. During the procedure, the surgeon finds that the mass is related to the ulnar nerve, takes a biopsy, and closes the wound.
- Pathological examination revealed a cellular schwannoma.
- Upon presentation to the author's institute, her examination revealed completely normal motor and sensory function of the ulnar nerve. Slightly positive Tinel's sign could be elicited along the sensory distribution of the ulnar nerve in the hand. An MRI of the right arm was suggestive of nerve sheath tumor (▶**Fig. 145.1**).
- Given the patient's younger age and the unpleasant cosmetic appearance, she elected to have the mass removed. A 10-cm incision was made to provide good proximal and distal exposure of the tumor (▶**Fig. 145.2**).
- Intraoperatively the tumor was related to the ulnar nerve and could be easily dissected off the nerve (▶**Fig. 145.3**). An entering and exiting small nerve fascicle was identified with no response on electrical stimulation (▶**Fig. 145.4**).
- The tumor was removed in one piece (▶**Fig. 145.5**). Upon follow-up, the patient continued to have normal ulnar nerve function with no motor or new sensory complaints.

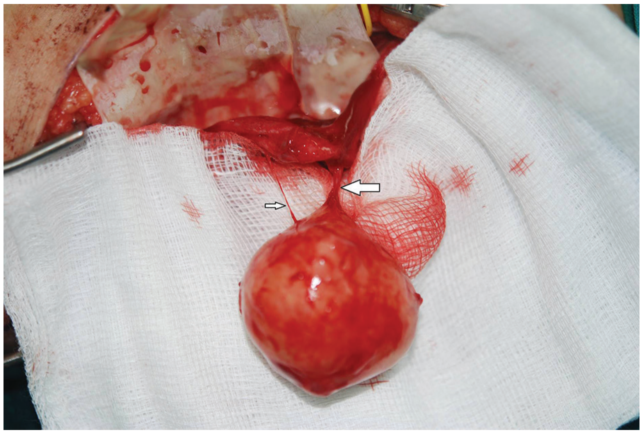

**Fig. 145.4** Following complete circumferential dissection of the mass, entering and exiting fascicles can be seen (*large* and *small arrows*, respectively), which can be divided.

7
8
9
10
11
12
13
14
15
3
4
6

**Fig. 145.5** It is a good practice to document the size of the resected mass with a picture using a reference ruler.

## Questions

1. Describe the MRI findings of benign nerve sheath tumors.
2. What are the common clinical presentations of nerve sheath tumors?
3. Classify nerve tumors.
4. What are the clinical and pathological differences between schwannomas and neurofibromas?
5. What is the role of biopsy in benign nerve sheath tumors?
6. What are the management options in benign nerve sheath tumors?
7. Describe the surgical strategies of benign nerve sheath tumors.

## Answers

1. *Describe the MRI findings of benign nerve sheath tumors.*
   - MRI is the radiological modality of choice in the diagnosis of benign nerve sheath tumors. The classical schwannomas will present as isodense to muscle on T1-weighted images and slightly hyperdense on T2-weighted images with the majority of lesions showing either dense or heterogeneous enhancement with contrast administration.
   - Malignancy is suspected if the lesions exceed 5 cm in diameter, exhibit peripheral enhancement or show perilesional edema.[1]
2. *What are the common clinical presentations of nerve sheath tumors?*
   - The majority of limb nerve sheath tumors present as a mass with no or mild neurological symptoms, mostly sensory. Body cavity nerve sheath tumors are detected radiologically as incidental findings when imaging is performed for nonrelated issues.
   - On examination the mass is usually firm, nontender, and mobile on the transverse axis of the nerve but not on the longitudinal axis. Positive Tinel's sign can be elicited when the mass is percussed.
   - Masses that presents with pain, immobility, and/or neurological deficits should raise the suspicion of malignant nerve sheath tumors as a diagnosis.[2,3,4]
3. *Classify nerve tumors.*[5]
   - Neoplastic
     - Nerve sheath origin
       - Benign
       - Schwannoma
       - Neurofibroma
       - Granular cell tumor
       - Perineurioma
       - Neurothekeoma
       - Malignant
       - Malignant peripheral nerve sheath tumor (MPNST)
     - Nerve cell origin
       - Primitive neuroectodermal tumor (PNET)
       - Neuroblastoma
       - Ganglioneuroma
       - Ganglioneuroblastoma
       - Chemodectoma
       - Pheochromocytoma
     - Metastatic tumors
   - Nonneoplastic
     - Intraneural lesions
       - Ganglion
       - Lipoma
       - Hemangioma
       - Desmoid

## Answers (*continued*)

  - Lipofibromatous hamartoma
  - Traumatic neuroma

4. *What are the clinical and pathological differences between schwannomas and neurofibromas?*
   - ▶**Table 145.1** summarizes the pathological differences between schwannomas and neurofibromas.
5. *What is the role of biopsy in benign nerve sheath tumors?*
   - When the clinical picture and the radiological appearance are highly suggestive of benign nerve sheath tumor, the benefit of the biopsy is not only small, but can be hazardous as it may cause nerve injury and/or neuropathic pain.
   - Biopsy is a reasonable option when the diagnosis of benign nerve sheath tumor is in doubt clinically or the radiological appearance suggests otherwise.[3]
6. *What are the management options in benign nerve sheath tumors?*
   - Observation and conservative treatment:
     - Asymptomatic patient with palpable mass only
     - Patient that are poor surgical candidates
   - Surgical resection:
     - Progressive neurological dysfunction
     - Pain
     - Local mass effect
     - Patient preference and cosmesis
     - Necessity for confirmation of tissue diagnosis in questionable cases
   - There is no clear role for biopsy, needle aspiration, or other adjuvant treatments such as radiation and chemotherapy.
7. *Describe the surgical strategies of benign nerve sheath tumors.*
   - When possible, the nerve sheath tumor exposure needs to be wide, this allow proximal and distal control and allows manipulation with possible rotation of the nerve and the tumor if needed.
   - Usually the tumor spreads the fascicles apart, and presents itself on the superficial aspect of the nerve, but occasionally the mass is hidden posteriorly and rotation of the nerve is required for safe removal of mass.
   - Once exposed, the surgeon needs to find a fascicle-free zone to access the tumor; this is done by incising multiple layers of connective tissue and the capsule of the nerve.
   - Adequate opening of the enclosing layers is evident by seeing a clear yellowish color which is characteristic of nerve sheath tumors.
   - Simple circumferential dissection is done till entering and exiting fascicles are identified. Both fascicles are divided and the mass is removed (provided the fascicles are not related to an important nerve function, as confirmed by intraoperative neuromonitoring stimulation).
   - In neurofibroma, more than one fascicle can be seen and the surgeon needs to make an intraoperative decision based on the clinical scenario and the results of the intraoperative stimulation to decide on the division of the fascicles.
   - It is essential to remember that the preservation of baseline motor function is the main objective of the surgery and leaving behind a small piece of tumor to preserve this function is wise in some cases.[6,7]

**Table 145.1** Clinical and pathological distinction of schwannomas and neurofibromas[5]

| Schwannoma | Neurofibroma |
|---|---|
| Typically affect extremities | Typically involves the trunk |
| May be associated with neurofibromatosis type 2 (NF2 associated) | Associated with neurofibromatosis type 1 |
| Nerve can often be identified with a single fascicle involvement | Nerve not usually identified as more than one fascicle involvement |
| Nerve is located along the periphery | Incorporates the nerve |
| Globular in shape | Varies in shape and form |
| May be cystic | Noncystic |
| Antoni A and B pattern | Uniphasic pattern |
| Mucin-poor matrix | Mucin-rich matrix |
| Palisading/Verocay bodies | Wagner–Meissner and occasional Pacinian-like corpuscles |
| Contains Schwann cells | Contains Schwann, perineurial, transitional cells, and fibroblasts |
| Typically no mast cells | Mast cells often present |
| Strong S-100 reactivity | S-100 reactivity in Schwann cell population, not in perineurial or fibroblasts |
| GFAP can be positive | Vimentin, CD 34 can be positive |

*Abbreviation:* GFAP, glial fibrillary acidic protein.

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# Case 146 Lower Extremity Peripheral Nerve Sheath Tumor

*Robert L. Tiel*

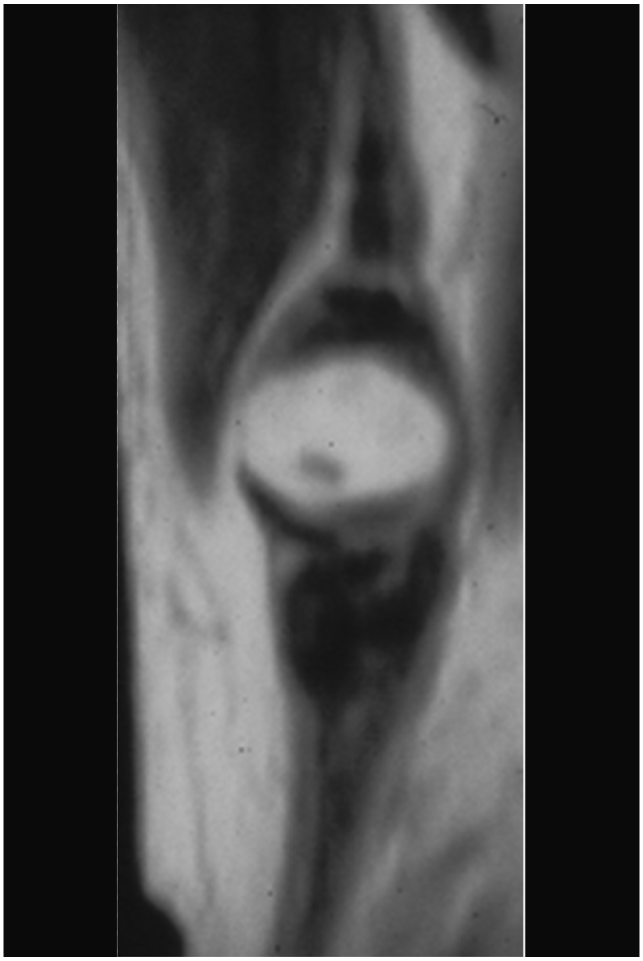

**Fig. 146.1** T1-weighted contrast-enhanced MRI of a schwannoma. Note widening of the nerve.

## Clinical Presentation

- A 15-year-old right-handed boy presents with a lump on the posterior lateral aspect of his right leg, just above the popliteal crease.
- Although the lump is usually painless, if knocked or manipulated he experiences a shooting sensation down his leg into the side and top of his foot.
- The frequent pains thus generated have forced him to abandon playing soccer.
- His neurologic examination is normal.
- When the lump is palpated it seems to have the dimensions of a large olive-sized (2 × 2 × 2 cm) mass. It moves from right to left, but not vertically. When lightly percussed, a sharp sensation is elicited which goes into the dorsum of the right foot.
- Other physicians had previously seen the patient. After an MRI scan had been obtained (▶**Fig. 146.1**), they advised him to have the lump monitored, to limit his activities, and to “live with” the condition.

## Questions

1. What is your working diagnosis?
2. Provide a detailed differential diagnosis.
3. What are the ancillary diagnoses for which he should be evaluated?
4. What are the clinical findings associated with these diagnoses?
5. The mother has a friend in whom neurofibromatosis (NF) was suspected; she states that this friend was sent to an ophthalmologist for diagnosis. Why?
6. She asks you if this tumor could be malignant. What is your answer?
7. The other physicians recommended "living with" the condition. What are this patient's options? What are the risks to the patient?
8. Should surgical excision be advised?
9. Should a needle biopsy be performed first?
10. Which incision should be used for general exposure?
11. Describe the steps in removing the tumor.
12. Should a frozen section be requested?
13. The tumor is removed; final sections are available for review 3 days later. Representative samples are shown in ▶**Fig. 146.2**, ▶**Fig. 146.3**, ▶**Fig. 146.4**, and ▶**Fig. 146.5**. What is your diagnosis?
14. Which immunohistochemical stains are useful in diagnosing peripheral nerve sheath tumors?

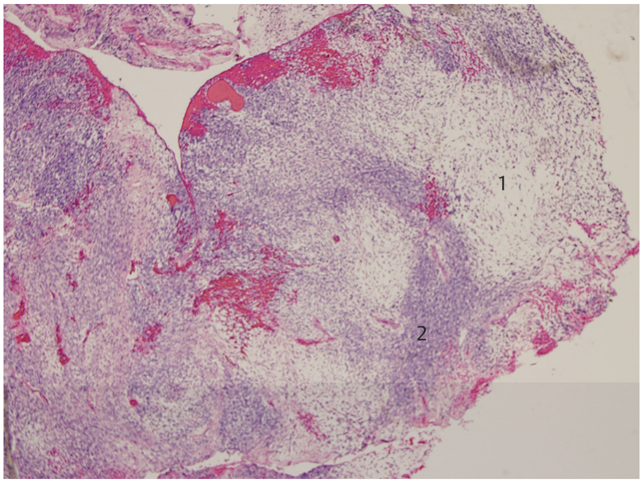

**Fig. 146.2** Low magnification (40×) of tumor with two areas of interest (hematoxylin and eosin stained).

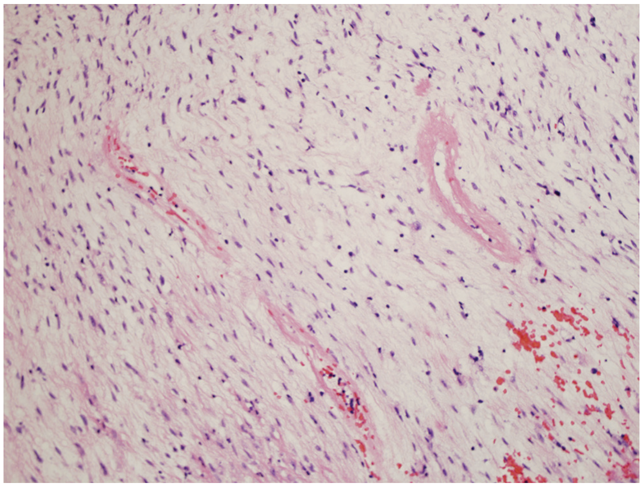

**Fig. 146.3** High magnification (200×) of area 1 (hematoxylin and eosin stained).

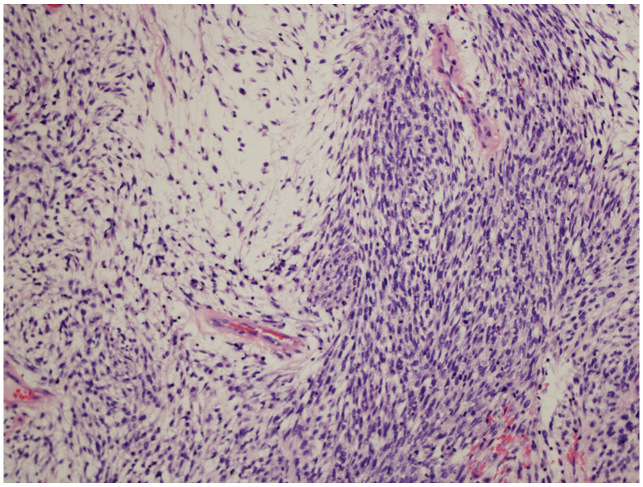

**Fig. 146.4** High magnification (200×) of area 2 (hematoxylin and eosin stained).

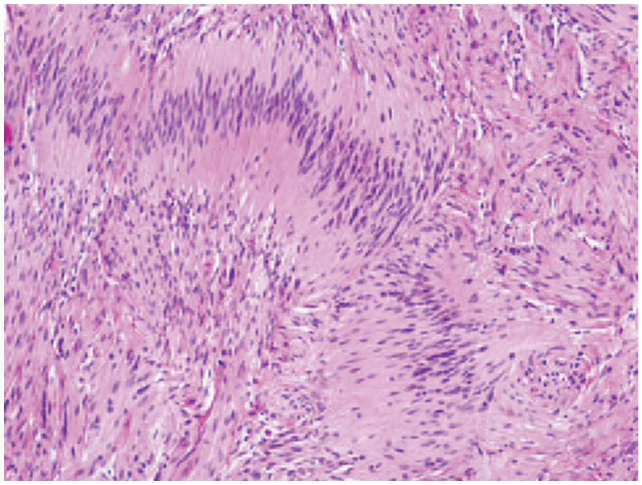

**Fig. 146.5** High magnification (200×) of tumor pathology (hematoxylin and eosin stained).

## Answers

1. *What is your working diagnosis?*
   - The working diagnosis is that of peripheral nerve sheath tumor.
   - The lump suggests tumor pathology. The MRI (▶**Fig. 146.1**) shows a contrast-enhancing intraneural mass widening the nerve.
   - The lateral mobility is evident, but vertical mobility strongly suggests a lesion within a movable peripheral nerve.
   - Finally, the paresthesias associated with manipulation point toward primary neural involvement in a specific peripheral nerve distribution (in this case the common peroneal nerve).
2. *Provide a detailed differential diagnosis.*
   The differential diagnosis of nerve sheath tumor includes the following[1]:
   - Benign tumors
     - Schwannoma
       - Cellular schwannoma
       - Plexiform schwannoma
       - Melanotic schwannoma
     - Neurofibroma
       - Diffuse (cutaneous)
       - Localized
       - Plexiform
     - Perineurioma
     - Nerve sheath myxoma
     - Granular cell tumor
     - Ganglioneuroma
   - Malignant tumors
     - Malignant peripheral nerve sheath tumors (MPNST)
     - Variants of above
     - Secondary neoplasms
   - Tumor-like lesions
     - Reactive lesions
       - Traumatic neuroma
       - Inflammatory pseudotumor
     - Inflammatory and infectious lesions
     - Intraneural ganglion cysts
     - Hyperplastic lesions
   - Localized hypertrophic neuropathy
     - Hamartomas
       - Fibrolipomatous hamartoma
3. *What are the ancillary diagnoses for which he should be evaluated?*
   - The presence of a peripheral nerve sheath tumor warrants evaluation for neurofibromatosis types 1 (NF-1) and 2 (NF-2) and for schwannomatosis.
4. *What are the clinical findings associated with these diagnoses?*
   - Diagnostic criteria for NF-1[2] (two or more of the following):
     - Six or more café-au-lait spots
       - Greater than 1.5 cm or larger in postpubertal individuals
       - Greater than 0.5 cm or larger in prepubertal individuals
     - Two or more neurofibromas of any type
     - One or more plexiform neurofibromas
     - Axillary or groin freckling, an optic pathway glioma, two or more Lisch nodules
     - A first-degree relative with NF-1 (as defined by the preceding criteria)
     - Characteristic osseous lesions, such as sphenoid dysplasia
   - Diagnostic criteria for NF-2[3]:
     - The main characteristic of NF-2 is the presence of bilateral vestibular schwannomas—tumors arising from the vestibular branch of the eighth cranial nerve (CN VIII).
     - Other diagnostic features consistent with NF-2:
       - A first-degree relative with NF-2
       - A unilateral vestibular schwannoma in a patient younger than age 30 years
     - Or any two of the following:
       - Meningioma
       - Glioma
       - Schwannoma
       - Juvenile posterior subcapsular lenticular opacities
       - Juvenile cortical cataracts
   - Diagnostic criteria for schwannomatosis[3,4]:
     - Individuals should not fulfill the diagnostic criteria for NF-2 or have any of the following:
       - A vestibular schwannoma of CN VIII
       - Constitutional NF-2 mutation
       - First-degree relative with NF-2
     - Definite schwannomatosis
       - Older than 30 years and two or more nonintradermal schwannomas (at least one confirmed by histology)
       - One schwannoma confirmed with histology and a first-degree relative who meets the above requirements
       - Lack of radiographic evidence of CN VIII tumor on an imaging study performed after age 18 years—possible schwannomatosis
       - Older than 30 years and two or more nonintradermal schwannomas (at least one confirmed by histology)
       - Older than 45 years and no symptoms of CN VIII dysfunction, and two or more nonintradermal schwannomas (at least one confirmed by histology)
       - Radiographic evidence of a schwannoma and a first-degree relative who meets the criteria for definite schwannomatosis
5. *The mother has a friend in whom neurofibromatosis (NF) was suspected; she states that this friend was sent to an ophthalmologist for diagnosis. Why?*

## Answers (*continued*)

- The ophthalmologists would be conducting a slit lamp evaluation to identify Lisch nodules or posterior subcapsular lenticular opacities.
- A Lisch nodule is a melanocytic hamartoma of the iris.
  - They appear after age 3 years.
  - They are present in 90% of NF-1 patients.
  - They are specific for NF-1.
  - They are usually clear yellow to brown.
  - A slit lamp examination may be necessary to differentiate them from nevi on the iris, where they present as flat or minimally elevated, densely pigmented lesions with blurred margins.
- Juvenile posterior subcapsular lenticular opacities
  - Usually asymptomatic
  - Occur in NF-2

6. *She asks you if this tumor could be malignant. What is your answer?*
   - The possibility of malignancy is always present until the permanent histologic sections are evaluated.
   - Incidence in the general population is 0.0001%.[5]
   - Although the likelihood of malignant transformation is higher in NF-1 patients, only half of malignant nerve sheath tumors arise from NF-1 patients; the other half arise de novo from people without NF.
   - From the clinical perspective, the tumor size and the presence of a more constant pain suggest the diagnosis of malignancy.
     - Loss of function in the distribution of the nerve suggests malignancy. Although benign nerve sheath tumors may attain a large size, they usually displace the fascicles aside and involve the fascicles minimally.
     - A Tinel's sign or mechanical irritability should not be confused with the pain of malignancy which is more constant and often throbs "at night."
7. *The other physicians recommended "living with" the condition. What are this patient's options? What are the risks to the patient?*

   The patient with a nerve sheath tumor has the following options:
   - Live with the tumor (observation)
     - Have its growth monitored
     - If it grows, have it operated upon
   - Have the tumor resected
     - Resection of the tumor causes little morbidity.
       - The risk of schwannoma together with a normal examination
   - Will lead to no postoperative motor deficit in 90% of operated cases
   - Resolves nerve irritability and pain in 75% of operated cases
     - The risk of solitary neurofibroma without motor deficit
   - Allows 78% of the patients to have no postoperative motor deficit
   - Allows pain to be resolved or improved in 88% of patients[6]
8. *Should surgical excision be advised?*
   - Given the relatively low risks of resection combined with the uncertainty of diagnosis, resection represents the single best option unless age or associated medical morbidity precludes surgery.
   - The lifetime focal cure rate for schwannomas is 95%.
   - The histologic diagnosis is determined and the tumor definitively treated.
9. *Should a needle biopsy be performed first?*
   - No, needle biopsy should not be advised for most nerve sheath tumors.[6]
   - In contradistinction to other soft-tissue tumors, biopsy of nerve sheath tumors increases the morbidity of management.
   - The biopsy needle must first blindly traverse the nerve sheath tumor, piercing the outside capsule and displaced fascicles, risking nerve damage to a functioning nerve.
   - Intraneural tissue planes that aid in tumor removal might be lost by biopsy-related hemorrhage or scarring.
   - An exception may be large painful tumors that show increased activity on positron emission tomography scanning because a targeted biopsy might allow determination of malignancy prior to operation.
10. *Which incision should be used for general exposure?*
    - An extensile incision should be located over the tumor and along the course of the nerve.
    - For the peroneal nerve above the popliteal fossa a "Lazy S" incision would be appropriate (▶ **Fig. 146.6**).
11. *Describe the steps in removing the tumor.*
    - The steps in tumor removal proximal to an entrapment site (for the peroneal nerve, the fibular head) include the following[7]:
      - The skin incision is made and the peroneal nerve and tumor are freed from all surrounding tissue.
      - In the location where the peroneal nerve can be entrapped by the fascial tissue at the head of the fibula, the nerve is "released" past the fibular head, well into the peroneus longus muscle.
      - Nerve action potential (NAP) recording (if available) is performed along the entire nerve. A NAP should be easily recordable (▶ **Fig. 146.7**).
      - Attention is turned to the tumor once the nerve and its branches have been completely neurolysed.
      - A fascicle-free area of the is tumor is determined
        - By inspection (▶ **Fig. 146.8**)
        - By cautious nerve stimulation (▶ **Fig. 146.9**)
      - Attempting to discover the dissection plane of the tumor, the surgeon makes an incision into the tumor and its primary fascicle and gentle dissection is used to develop this plane between tumor and fascicles (▶ **Fig. 146.10**).

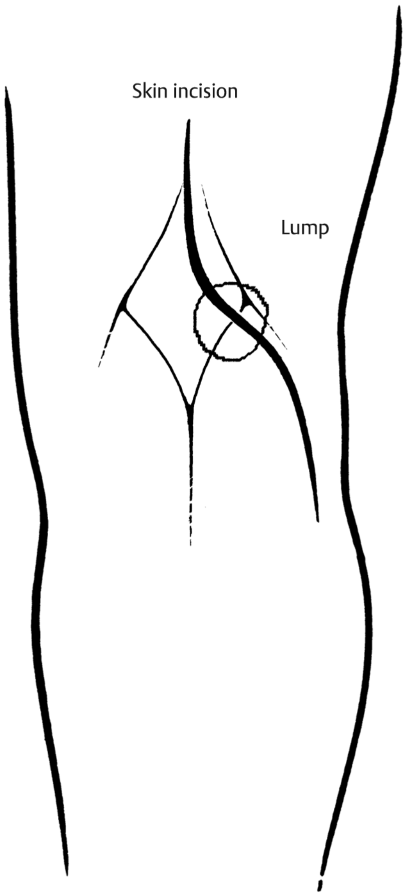

**Fig. 146.6** Incision for exposing the right common peroneal nerve to expose the nerve sheath tumor.

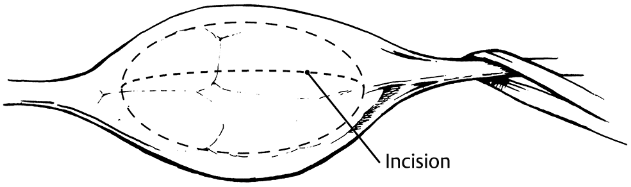

**Fig. 146.8** A fascicle-free area is determined from inspection.

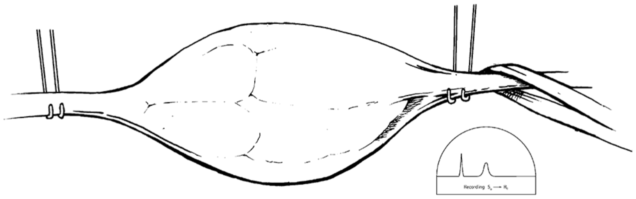

**Fig. 146.7** The tumor and nerve are dissected free from surrounding connective tissue and a positive nerve action potential is recorded.

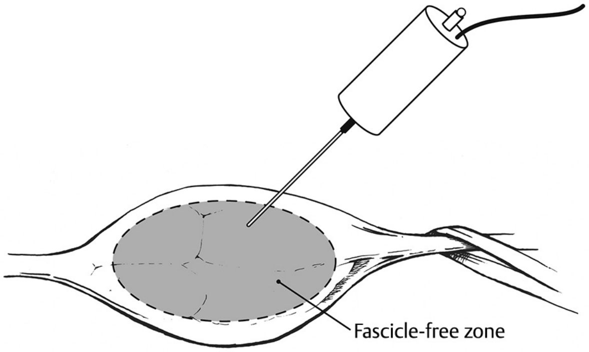

**Fig. 146.9** An area of no stimulation is mapped out using a disposable hand-held stimulator set on 2 mA.

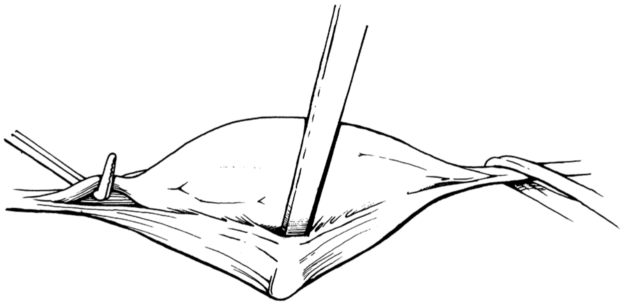

**Fig. 146.10** An incision is made into the nerve and a plane of dissection between the tumor and the nerve is developed.

## Answers (*continued*)

  - The tumor and its primary entry and exit fascicle are dissected out (▶ **Fig. 146.11**).
  - A NAP recording is done of the entering fascicle (▶ **Fig. 146.12**). A flat trace is elicited, thereby ensuring that the fascicle from which the tumor arose is nonfunctioning or solely sensory.
  - The tumor is removed well into the originating and exiting fascicle to ensure complete tumor excision (▶ **Fig. 146.13**).
  - A whole nerve NAP is recorded after resection to insure a functioning nerve.
  - The meticulous hemostasis of the tumor bed is achieved prior to closure.
  - The incision is closed.

12. *Should a frozen section be requested?*
    - A frozen section is usually unnecessary if the tumor dissection proceeds easily.
    - If dissection planes are not found:
      - The nerve adheres to soft-tissue structures, and a frozen section may be warranted to evaluate the suspected tissue for malignancy or alternative diagnosis.
    - If the nerve seems unusually hard or fibrotic:
      - The diagnosis of peripheral nerve pseudotumor is now being considered, and a representative sample is usually warranted to determine whether dissection should proceed.
13. *The tumor is removed; final sections are available for review 3 days later. Representative samples are shown in* ▶ **Fig. 146.2**, ▶ **Fig. 146.3**, ▶ **Fig. 146.4**, *and* ▶ **Fig. 146.5**. *What is your diagnosis?*
    - The diagnosis is benign schwannoma.[8]
    - Antoni A pattern (▶ **Fig. 146.4**)
      - Compact elongated cells with tapered spindle-shaped nuclei
      - Variable chromasia
      - Ample pink cytoplasm
    - Antoni B pattern (▶ **Fig. 146.3**)
      - Loose texture “cobweb-like meshwork”
      - Multipolar processes
      - Round and oval nuclei
      - Occasional microcysts
    - Verocay bodies (▶ **Fig. 146.5**)
      - Nuclear clusters in a palisading arrangement
      - Double rows of nuclei separated by aligned eosinophilic cell processes (▶ **Fig. 146.14**)
14. *Which immunohistochemical stains are useful in diagnosing peripheral nerve sheath tumors?*
    - The immunohistologic stains useful in identifying peripheral nerve tumors are summarized in **Table 146.1**.[9–14]
    - They include:
      - S-100 (so called because of its solubility in 100% saturated ammonium sulfate)
      - Cluster of differentiation molecule (CD-34)
      - Epithelial membrane antigen (EMA)
      - Nestin

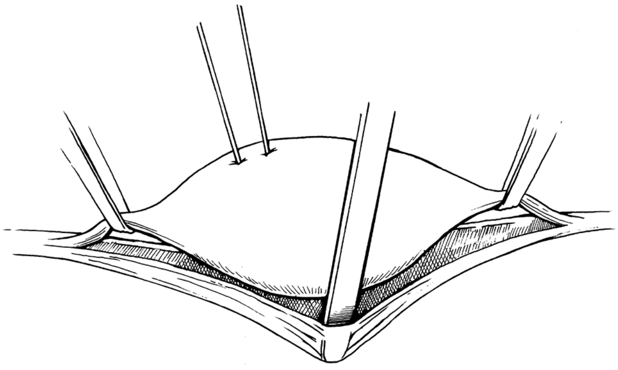

**Fig. 146.11** Stitch retraction is sometimes useful in manipulating the tumor to aid in the intraneural dissection of the tumor from the surrounding nerve.

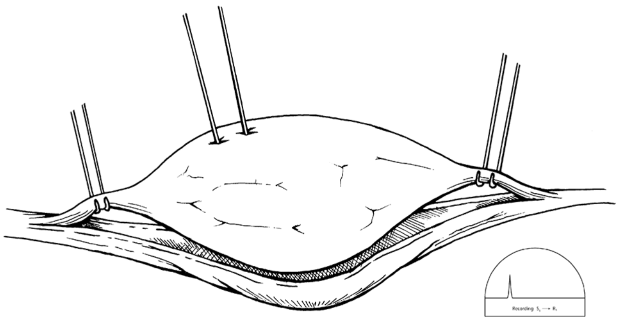

**Fig. 146.12** A nerve action potential cannot be recorded across the involved fascicle indicating that the fascicle is either pure sensory or contains no useful motor fibers.

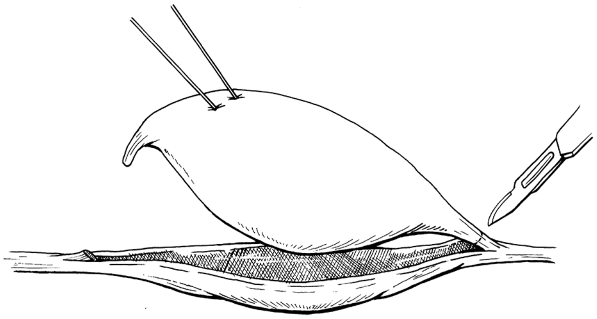

**Fig. 146.13** The tumor is sharply cut at both ends where the fascicle of origin is of normal caliber.

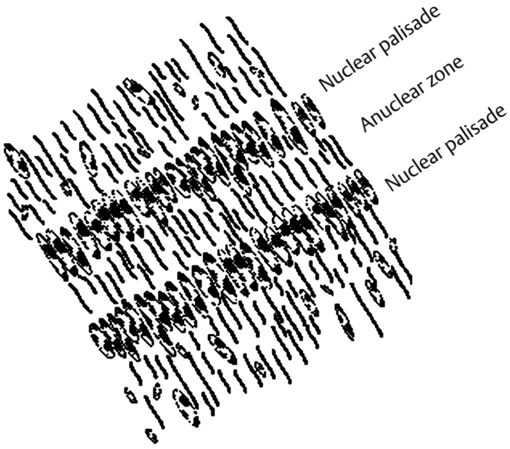

**Fig. 146.14** Diagram of a Verocay body.

**Table 146.1** Immunohistochemistry of peripheral nerve sheath tumors

| Marker | Function | Specificity | Schwannoma | Neurofibroma | Perineurioma[9–11] | MPNST |
|---|---|---|---|---|---|---|
| S-100 | Described initially by Moore,[12] constitutes a large family of at least 20 proteins with calcium binding ability | Present in the cytosol of glial and Schwann cells, and also in adipocytes and chondrocytes, although in very low concentrations in the latter two | Strongly positive ++++ | Variable ++ to ++++ | Less than 5% positive − to + | Variable 50% positive − to ++ |
| CD-34 (human progenitor cell antigen)[13] | | Immunoreactive to endoneurial fibroblasts | Negative Antoni A, positive Antoni B − to + | Positive +++ | Variable − to ++ | Variable − to + |
| EMA[10,14] | A large, highly glycosylated protein (MUC1) | Expressed on the apical membrane of many epithelial cells | Negative − | Variable − to + | Positive ++++ | Variable − to + |
| Nestin[11] | Intermediate filament protein | Expressed in neuroectodermal stem cells | Variable − to ++ | Variable − to + | | Positive +++ |

*Abbreviations:* +, positive expressivity; −, negative expressivity; EMA, epithelial membrane antigen; MPNST, malignant peripheral nerve sheath tumor.

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# Case 147 Foot Drop and Peroneal Nerve Injury

*Robert L. Tiel*

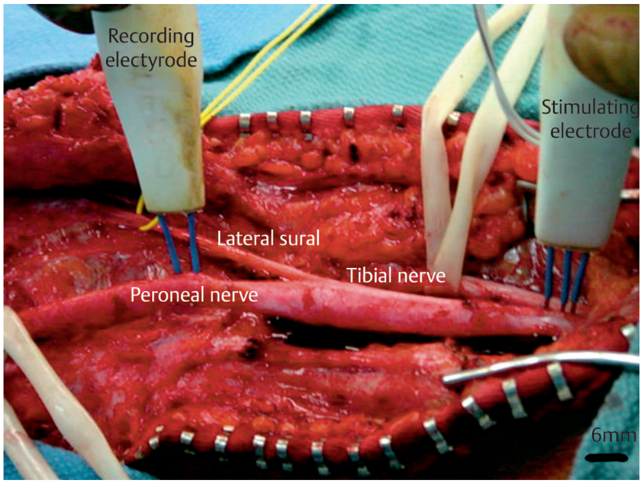

**Fig. 147.1** Nerve action potential recording of a stretched peroneal nerve showing increased diameter well into the sciatic nerve at the sciatic bifurcation into the common peroneal nerve and the tibial nerve.

## Clinical Presentation

- An 18-year-old college freshman suffers a right knee dislocation during fall football practice.
- He has immediate loss of dorsiflexion of the right foot on the field.
- He is discovered to have an anterior and posterior cruciate ligament (ACL; PCL) and lateral collateral ligament tears.
- He is scheduled for orthopaedic knee surgery and his ligaments are repaired.
- The peroneal nerve, which was seen in surgery, was believed to be "intact" but slightly bruised.
- He is seen 4 months later in consultation with the following physical examination of his right leg.
  - Motor examination right lower extremity (Medical Research Council scale)
    - Iliopsoas: 5/5
    - Quadriceps femoris: 5/5
    - Tibialis anterior: 0/5
    - Extensor hallucis longus: 0/5
    - Extensor digitorum communis: 0/5
    - Peroneus longus/brevis: 3/5
    - Posterior tibialis: 4+/5
    - Plantar flexion (soleus/gastrocnemius): 4+/5
    - Toe flexion: 4+/5
  - Sensory examination of the right lower extremity
    - Web space between first and second toes numb to pin
    - Dorsum of foot numb to pin
    - Lateral border of foot slightly decreased compared with left foot
    - Sole of foot same compared with the left foot
  - Deep tendon reflexes (DTRs)
    - Right 2+ knee jerk (KJ) 1+ ankle jerk (AJ) (with reinforcement)
    - Left 2+ KJ 2+ AJ

## Questions

1. What physical finding must be checked on the football field and in the emergency room?
2. Is the nerve injured?
3. What is the Seddon or Sunderland grade?
4. Where is the nerve(s) injured?
5. Does the patient need any orthosis? If so what type?
6. Which plan of care should be arranged?
7. Which electrodiagnostic tests should be ordered and when? Which muscles should be examined?
8. When, if ever, should this nerve injury be explored?

## Questions (*continued*)

9. What incision should be used? How do you find the peroneal nerve when all you see is fat? What is the relation of the common peroneal nerve to the fibular head?
10. When, if ever, should the nerve(s) be repaired?
11. What are the results of operative repair at this level for foot drop?
12. What are patient's alternatives to nerve repair?

## Answers

1. *What physical finding must be checked on the football field and in the emergency room?*
   - The most important physical finding in this case was the presence of a palpable distal pulse.
   - Unrecognized ischemia can lead to loss of limb and even death.
   - Using the Doppler ultrasound and/or angiography to assess the vasculature can further ensure that the leg is viable.
2. *Is the nerve injured?*
   - Yes, there is no function of the muscles of dorsiflexion and no mention of direct injury to the muscles; therefore, the nerve is injured and not working properly after injury.
3. *What is the Seddon or Sunderland grade?*
   - The injury is either axonotmetic or neurotmetic by the Seddon classification or Sunderland grade 2–4 (▶**Table 147.1**).
   - Determining either the Seddon or Sunderland grade acutely is impossible upon initial presentation.
   - Because after 4 months, there has been no recovery of function, then this is not a neurapraxic injury—not a (Seddon) or a Sunderland grade 1 injury.
   - The Sunderland grade cannot be 5 as the nerve was inspected and appears to be in continuity.
4. *Where is the nerve(s) injured?*
   - Localization of the nerve injury is at the level of the peroneal division from the sciatic nerve.
   - There is mild injury of the tibial division and complete injury of the peroneal.
   - The peroneal nerve is tethered at this bifurcation and some damaging energy has been transmitted to the tibial nerve.
   - The normal cross-sectional diameter of the sciatic nerve just proximal to the split is 11 to 12 mm, the tibial nerve is usually 6 mm and the peroneal nerve 5 mm at its origin.
   - After a stretch injury, these measurements increase and the peroneal nerve might be 11 to 12 mm and the tibial nerve 7 mm demonstrating the internal fibrosis and injury which accompanied the dislocation (▶**Fig. 147.1**).
5. *Does the patient need any orthosis? If so what type?*
   - Almost all patients will benefit from a well-fitting orthosis.
   - The most common is the in the shoe ankle foot orthosis (AFO), but there exist many variations on this model. The specific purpose of an AFO is to provide toe dorsiflexion during the swing phase, medial and/or lateral stability at the ankle while standing, and, if necessary, push-off stimulation during the late stance phase.
   - The goal of bracing is to prevent toe and foot drop while walking, which would contribute to tripping.
   - In addition, the ankle also needs the lateral support to avoid spontaneous inversion and twisting of the ankle joint.
   - For those so inclined, a stiff lace-up work boot or a cowboy boot can reasonably serve these dual purposes.
6. *Which plan of care should be arranged?*
   - The essential question with all nerve injuries and this one in particular is whether the nerve will recover spontaneously or not.
   - Waiting 3 to 4 months is recommended unless evidence exists that the nerve is divided.
   - Fitting for an orthosis and treating symptomatically for pain are both interventions usually warranted.
7. *Which electrodiagnostic tests should be ordered and when? Which muscles should be examined?*
   - Waiting 4 months, all hope of a neurapraxic recovery has disappeared.
   - An electromyographic (EMG) study is done to determine whether any nerve fibers have returned to their respective muscles through the injury.
   - An axonotmetic injury (Sunderland 2) will usually recover on its own.
   - The pattern of injury indicates that both the tibial and peroneal divisions of the sciatic nerve have been involved.
   - The goal of EMG is to determine the range of involvement. The short head of biceps is supplied by the lateral peroneal division of the sciatic nerve. A normal pattern here ensures that the injury is lower on the sciatic nerve.
   - Muscles innervated by the peroneal nerve should be examined. As the peroneal nerve divides into the superficial peroneal nerve and the deep peroneal nerve, representative muscles from both peroneal divisions should be examined.
   - These muscles include the tibialis anterior from the deep peroneal nerve and peroneus longus from the superficial peroneal nerve.

## Answers (*continued*)

- Tibial nerve-innervated muscles should also be evaluated, such as the gastrocnemius muscle and the posterior tibial muscle.
- EMG involvement of the posterior tibial muscle in cases of foot drop will localize the injury to a level at or above the sciatic nerve bifurcation.
- An isolated peroneal injury will not show involvement of the posterior tibial muscle.

8. *When, if ever, should this nerve injury be explored?*
   - The nerve should be explored 3 to 4 months after injury if no recovery is observed and none demonstrated on EMG.[1]
   - If EMG shows some recovery, it is worth waiting 8 weeks and repeating the clinical and, if necessary, electrical examination.
   - When EMG recovery is not enough to power useful regeneration, then operative repair is required.
9. *What incision should be used? How do you find the peroneal nerve when all you see is fat? What is the relation of the common peroneal nerve to the fibular head?*
   - A "lazy S" incision on the back of the leg provides adequate "extensile" exposure and avoids going perpendicular to the knee flexion crease (▶**Fig. 147.2**).
   - The tibial and peroneal nerves split from the sciatic at the lower third of the thigh (▶**Fig. 147.3**).
   - The peroneal nerve lies along the medial edge of the short head and medial edge of the tendon of the biceps femoris, which is easily differentiated from the surrounding fat (▶**Fig. 147.4**).
   - The peroneal nerve can be palpated at the fibular head.
   - This is a point of fixation where the common peroneal nerve dives under the peroneus longus muscle.[2]
10. *When, if ever, should the nerve(s) be repaired?*
   - The nerve should be repaired when no clinical or EMG evidence of recovery exists.
   - Nerve action potential (NAP) recording can be done over the affected segment (▶**Fig. 147.1**).
   - If a regenerative response is noted, then spontaneous recovery to M3 or greater will occur 90% of the time.
   - If no recording is elicited, the damaged segment will be resected back to healthy, bleeding, and

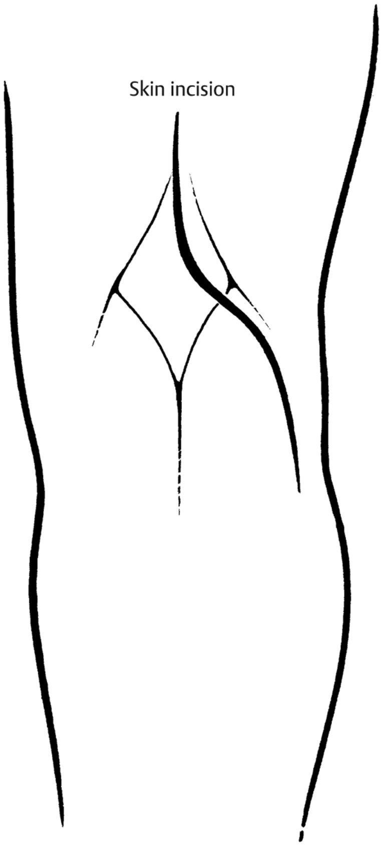

**Fig. 147.2** The "lazy S" incision used for peroneal nerve exposure of the right peroneal nerve, posterior view.

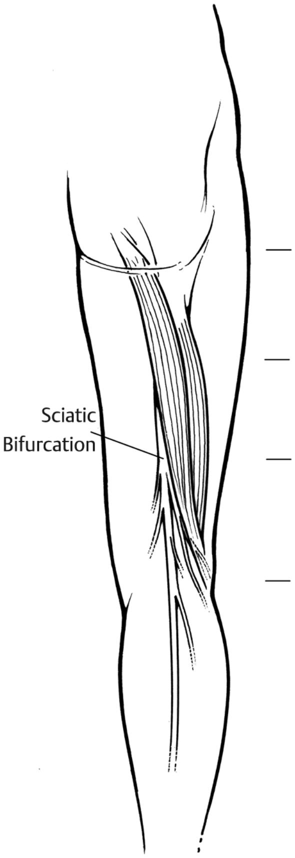

**Fig. 147.3** The thigh can be divided into thirds. The bifurcation of the sciatic is at the junction of the lower and middle third of the thigh.

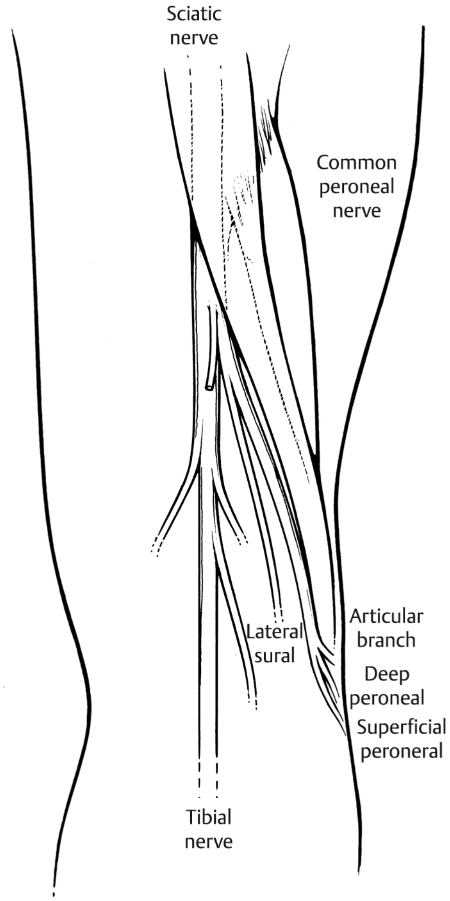

**Fig. 147.4** The peroneal nerve is to be found on the medial side of the junction of the short head of biceps and the long head of the biceps femoris tendon.

## ■ Answers (*continued*)

pouting fascicles and the gap repaired with sural nerve grafts.

- If the damaged segment is greater than 10 cm, orthopaedic procedures of recovery with graft repair are so poor that closure may be all that is indicated at this time. An orthopaedic procedure such as tendon transfer may then be contemplated at a later time.

11. *What are the results of operative repair at this level for foot drop?*
    - The results of operative repair for peroneal nerve injury are very much distance dependent.
    - Pooled literature shows a 29 to 50% recovery to M3 or greater with graft repair and a 39 to 100% recovery with primary suture repair.[3]
    - Graft length is a determinate with 70% recovery for grafts < 5 cm, 35% for grafts 6 to 12 cm and 18% for grafts over 12 cm.[4]
    - Most knee dislocations injure the nerve from its fibular fixation into the sciatic bifurcation, thereby requiring long grafts, usually > 12 cm, thus predicting poor results.
12. *What are patient's alternatives to nerve repair?*
    - Currently, no acceptable nerve transfers for this nerve injury exist.
    - After a year without neurologic recovery, orthopaedic solutions become necessary.
    - The posterior tibialis tendon provides the energy of dorsiflexion.
    - Depending upon the completeness of injury, different variations of tendon transfer are employed.[5]
    - After minor recovery in the evertor muscles (M2), tibialis posterior transfer appears to be more effective.

**Table 147.1** Correlation between seddon and sunderland nerve injury grading

| Clinical State of the Nerve | Demyelination, Conduction Block | Disruption of the Nerve Fibers, Endoneurium Intact | Disruption of the Endoneurium, Perineurium Intact | Disruption of the Perineurium, Epineurium Intact | Complete Disruption of the Epineurium, Nerve Severed |
|---|---|---|---|---|---|
| Seddon | Neuropraxia | Axonotmesis | Neurotmesis | Neurotmesis | Neurotmesis |
| Sunderland | Grade 1 | Grade 2 | Grade 3 | Grade 4 | Grade 5 |

*Source:* Seddon H. Surgical Disorders of the Peripheral Nerves. Edinburgh: Churchill Livingstone; 1972; Sunderland S. Nerve and Nerve Injuries. New York: Churchill Livingston; 1978.

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# Case 148 Gunshot Injury to the Sciatic Nerve

*Bassam M. J. Addas*

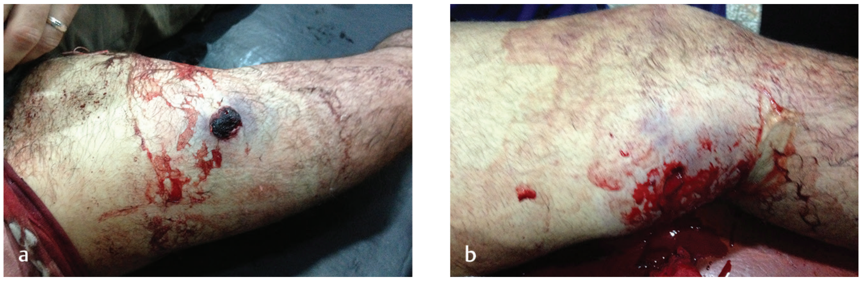

**Fig. 148.1** Photographs demonstrating the bullet entry (**a**) and exit (**b**) wounds. Note the swelling of the thigh which represent an underlying hematoma.

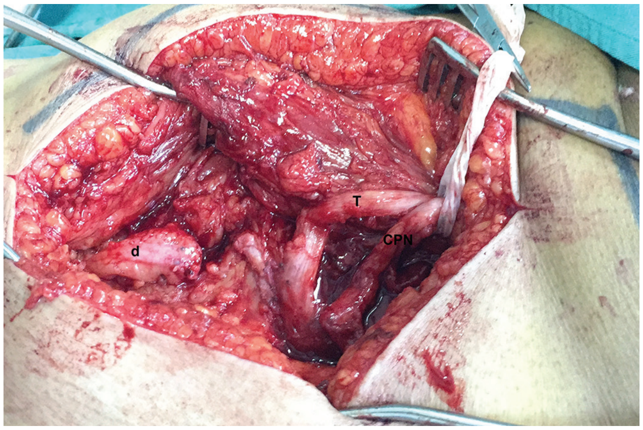

**Fig. 148.2** Surgical photograph following exposure of the sciatic nerve showing the proximal end as well as both distal ends: the tibial (T) and common peroneal (CPN) divisions, which are scarred down in the surrounding soft tissue with complete disconnection from the proximal stump (d).

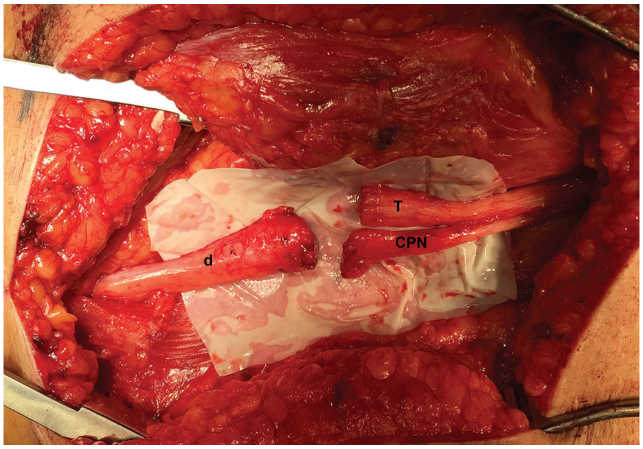

**Fig. 148.3** Following external neurolysis of the proximal and distal stump from the surrounding tissues, both tibial (T) and common peroneal (CPN) divisions were neurolysed in preparation for the repair. The proximal stump (d) will be neurolysed to its division prior to repair.

## Clinical Presentation

- A 27-year-old male suffered a gunshot wound (GSW) to lateral aspect of the right thigh.
- In the emergency room, the patient was found to have stable vital signs. The main complains were of numbness along his foot and inability to move his ankle in both directions (flail foot).
- Clinical examination of the right lower limb revealed puncture wounds along the lateral and medial aspects of the thigh, representing the entry and exit of the bullet (▸**Fig. 148.1**). The patient was unable to perform both dorsi- and planter flexions of the ankle, and had no sensation along the dorsum and the sole of the foot, except from a small area in the medial aspect of the middle of the foot.
- The diagnosis of complete sciatic nerve palsy was entertained. The patient was observed for about 6 months with no recovery, and it was subsequently decided to explore the injured area with the goal of primary end-to-end repair of the sciatic nerve (▸**Fig. 148.2**, ▸**Fig. 148.3**, and ▸**Fig. 148.4**)

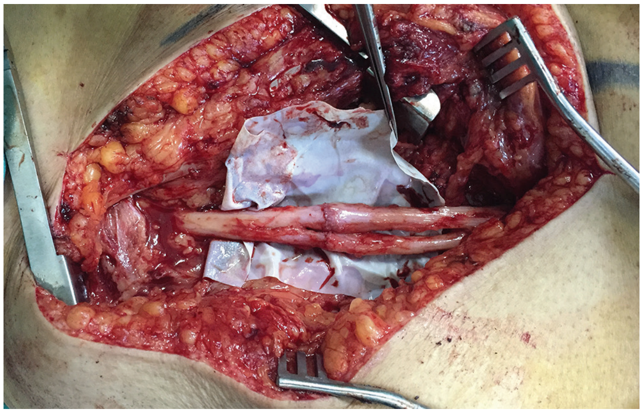

**Fig. 148.4** Following proximal stump separation into both divisions, individual repair of both divisions of the sciatic nerve was performed using 8/0 nylon suture under loupe magnification.

## Questions

1. How would you clinically assess a patient with possible acute injury of the sciatic nerve?
2. List the important muscles supplied by each branch of the sciatic nerve and its major function and describe its sensory supply.
3. What is the natural history of patients with GSW injuries to the peripheral nerves?
4. What are the management options of patients of GSWs to the sciatic nerve?
5. What are the factors that influence the results of nerve repair?

## Answers

1. *How would you clinically assess a patient with possible acute injury of the sciatic nerve?*
   - It is well known that GSW injuries to the peripheral nerves do not necessarily cause a complete transection of the nerve, yet it is not surprising that we find initially a picture of complete palsy, which is usually attributed to contusion of the nerve, compression by hematoma or surrounding tissues, and nerve edema.
   - The sciatic nerve terminates into common peroneal and tibial branches. The common peroneal nerve supplies the peroneal muscles (ankle evertors) and the tibialis anterior (ankle dorsiflexor). The tibial nerve supplies the gastrocnemius (ankle plantar flexors).
   - Inability to perform dorsiflexion and plantarflexion is the hallmark of sciatic nerve dysfunction. This is a very handy and quick way to evaluate sciatic nerve function in the emergency setting.
2. *List the important muscles supplied by each branch of the sciatic nerve and its major function and describe its sensory supply.*

   The sciatic nerve can be considered as two separate nerves (tibial and common peroneal) rather than a single nerve with two branches, as they share a common epineural sheath from its emergence from the pelvis through the greater sciatic notch.[1] However, it is commonly considered as one nerve all the way down to the lower third of the thigh, where it bifurcates into the tibial and common peroneal divisions. Giving this information, we can list the branches as:

   - Sciatic nerve proper (before the anatomical bifurcation) with innervation of:
     - Hamstrings (biceps femoris, semitendinosus, and semimembranosus) which are the major knee flexors
   - Tibial nerve with innervation of:
     - Gastrocnemius (lateral and medial heads), major ankle flexor
     - Soleus (ankle flexor)
     - Tibialis posterior (chief ankle invertor)
     - Flexor hallucis longus (big toe flexor)
     - Flexor digitorum (toe flexor)
     - Foot muscular layers (four layers)

       The tibial nerve sensory distribution is along the sole of the foot with the exception of a small island on the medial aspect of the middle of the foot which is innervated by the saphenous nerve. A small area behind the lateral malleolus is innervated by the sural nerve.
   - Common peroneal nerve
     - Superficial peroneal nerve with innervation of:
       - Peroneus longus (ankle evertor)
       - Peroneus brevis (ankle evertor)

         The superficial peroneal nerve sensory

## Answers (*continued*)

distribution is along the anterolateral aspect of the leg with the dorsum of the foot excluding the first web space which is innervated by the deep peroneal nerve.

  - Deep peroneal nerve with innervation of:
    - Tibialis anterior (ankle dorsiflexor)
    - Extensor hallucis longus (big toe extensor)
    - Extensor digitorum (toes extensor)
- See ▶ **Fig. 148.5** for illustrated course.

3. *What is the natural history of patients with GSW injuries to the peripheral nerves?*
   - The natural history of isolated GSW to the sciatic nerve is generally fair as one-third of patients will recover spontaneously and two-thirds will requires surgical exploration, of those one-third will benefit from external neurolysis alone, and only one-third will require primary or graft repair.[2–4]
   - When the injury is complicated by long bone fractures, vascular injuries, and soft tissue loss, the chances of good recovery will drop.
4. *What are the management options of patients of GSWs to the sciatic nerve?*
   - The surgeon is faced with two scenarios, either conservative management or surgical intervention.[2,5,6,7] This depends mainly on the recovery of the target muscles.
   - When the target muscle does not show any recovery in the first 6 months, surgical exploration is offered.

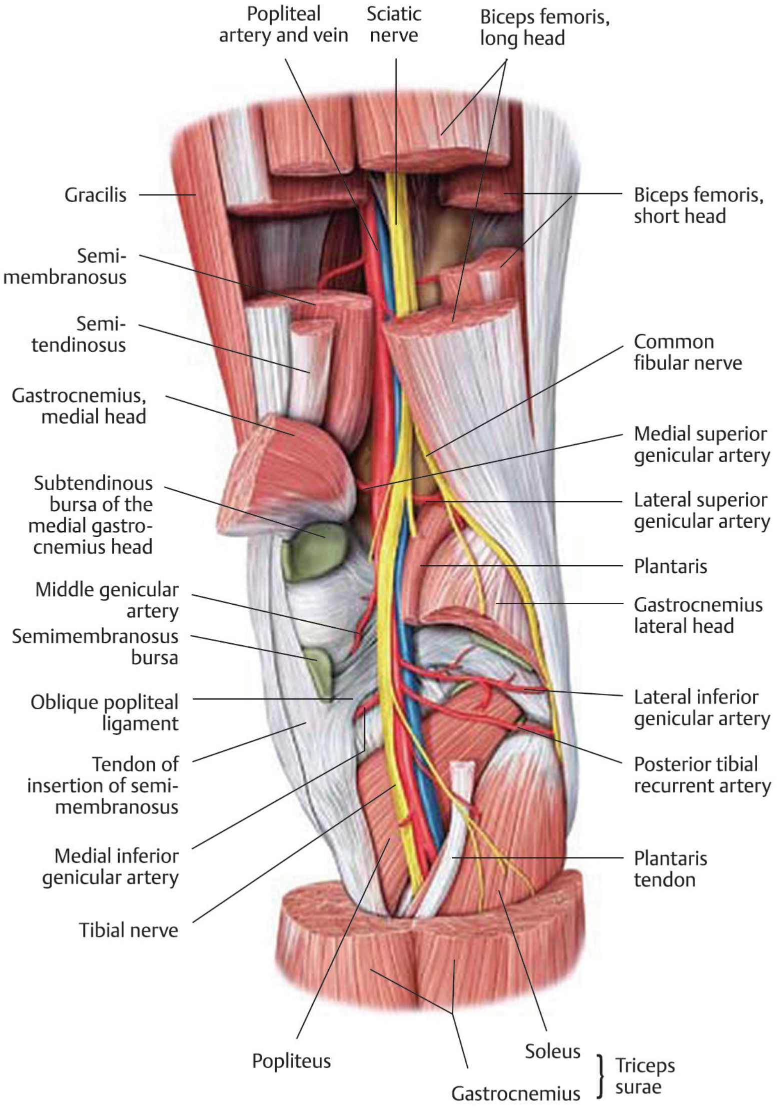
Popliteal artery and vein
Sciatic nerve
Biceps femoris, long head
Gracilis
Biceps femoris, short head
Semi-membranosus
Semi-tendinosus
Common fibular nerve
Gastrocnemius, medial head
Medial superior genicular artery
Subtendinous bursa of the medial gastro-cnemius head
Lateral superior genicular artery
Plantaris
Middle genicular artery
Gastrocnemius lateral head
Semimembranosus bursa
Lateral inferior genicular artery
Oblique popliteal ligament
Tendon of insertion of semi-membranosus
Posterior tibial recurrent artery
Medial inferior genicular artery
Plantaris tendon
Tibial nerve
Popliteus
Soleus
Gastrocnemius
Triceps surae

**Fig. 148.5** Detailed anatomic rendering of the course of the sciatic nerve in the popliteal fossa. Note that "peroneal" and "fibular" nerves are used interchangeably. Innervated muscles are also identified and labeled. (Reproduced with permission from Schuenke M. et al. Thieme Atlas General Anatomy and Musculoskeletal System. New York: Thieme; 2010: 501; Illustration by Karl Wesker/Markus Voll.)

## Answers (*continued*)

- In cases of associated femur fracture with nonhealing or the presence of external fixation, surgery should be delayed until complete bone healing takes place.

5. *What are the factors that influence the results of nerve repair?*
   - The level of the nerve injury (the further the target muscle to be innervated by the injured nerve, the worse the prognosis). Intrinsic foot muscle in this case will be the last to recover.
   - The delay from time of injury to time of repair (generally a conservative period of 4 to 6 months is accepted). Delayed repair or more than 1 year's wait will generally give poor results due to the lysis of the neuromuscular junction.
   - Primary nerve repair is generally superior to graft repair (single suture line versus two suture lines).
   - Associated soft tissue loss, long bone fracture, and vascular injuries will generally complicate the healing of repaired nerves.
   - Advanced age is considered a negative factor in nerve healing and the age of 40 years and older is usually associated with poor results.[2,8]

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